EARLY DIAGNOSIS

BY VARIOUS AUTHORS

EDITED BY

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PREFACE

THE early recognition of important disease is the special responsibility of the general practitioner for whom this book has been written It is he who must appreciate the signs which will lead him to commission on behalf of his patients the elaborate expensive and sometimes even potentially dangerous machinery of modern medical investigation Not only a source of intellectual satisfaction and the basis of reliable prognosis early diagnosis has been endowed by the headlong advance of therapeutics with a practical importance undreamed of even a few years ago No doubt the near future will furnish further instances such as those of bacterial endocarditis and tuberculous meningitis in which early diagnosis may already mean for the patient literally the difference between life and death

For these reasons no apology is offered for the presentation of these practical essays dealing with an aspect of clinical medicine often covered only by implication in the standard texts

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CHAPTER 1

ON EARLY DIAGNOSIS

BY LORD COHEN OF BIRKENHEAD

THE **purpose of diagnosis is action,** the purpose of **early diagnosis** is to institute as early as possible all measures which may be indicated for the cure alleviation and prevention of complications of a patient s illness and for the protection of others

The method of diagnosis is the same whether the patient is seen early or late in the course of his disease Occasionally the intelligent knowledgeable and experienced doctor is able to make a spot diagnosis for he is familiar with the face of disease and even the medical novitiate should discern the blatant stigmata of Graves disease or myxoedema of measles or chickenpox of rheumatoid arthritis or scleroderma Not even the most expert physician however can afford to dispense with that method of diagnosis hallowed by custom especially in the early phases of disease for here classical signs may not have developed In early thyrotoxicosis there may be neither exophthalmos nor goitre and in Parkinsonism neither paralysis nor tremor Moreover the overt diagnosis *e g* of Graves disease may not be complete bacterial endocarditis or pulmonary tuberculosis may be associated and must be recognized if life is to be preserved

The method in its simplest terms is based on *eliciting a history* and *making an examination*

The patient s story (and that of relatives and friends where necessary) must be listened to attentively for more often than not this will give the diagnosis What are the symptoms? (and the doctor must not be beguiled by such words as wind — which may mean a coronary occlusion or rheumatism —which may mask tabes dorsalis he must seek to elicit in detail and to

understand the patient s complaints in language which means the same both to doctor and patient) How long have they troubled him? In what order did they appear? How have they varied during the course of the illness? Do the past or family history occupation habits or other factors appear to bear on the illness? Clearly the overgarrulous must be curbed but high amongst the qualities of the good diagnostician is his ability rapidly and accurately to elicit discern and enquire directly about salient points in the history

In many diseases the history alone points to the diagnosis when even on the most painstaking physical examination no abnormalities can be found or signs are few and equivocal Thus the history of recurring episodes of transient pareses paraesthesiae blindness or vertigo may be the only yet the firmest evidence of disseminated sclerosis Again in most diseases when the examination is negative it is the history which points to the need for special investigations Thus anorexia wasting and epigastric discomfort may be the only pointers to a large cancer of the stomach which is not palpable but only too evident on X ray or cough and haemoptysis the only hints of a cancer of the bronchus easily seen on bronchoscopy but giving no physical or X ray signs of its presence

Then comes the **examination** by inspection palpation percussion and auscultation—never to be neglected but even more meticulously to be followed in the early stages of disease for the less obvious the signs the more assiduously must they be sought How often is Parkinsonism overlooked or labelled rheumatism or writer s cramp because the absence of the normal swing of the arm on walking has not been noticed? Or more grievous offence how often is a cancer of the colon or rectum missed in an operable stage because a diagnosis of constipation has satisfied and no abdominal or rectal examination has been made?

The examination reveals the condition of the patient at a given moment of time The history tells of events which have led to the present condition of the patient and seeks to elucidate it But the accuracy of a history depends on the patient s memory and veracity Thus *when the history appears to conflict with*

physical findings on examination the history is likely to be the less reliable For example many a woman with a cancer of the breast fixed to skin and chest wall will give a history of having noticed a lump only a day or two before

Re-examination at intervals of a few hours days or weeks depending on the acuteness of the illness is of the greatest value in diagnosis for the changes which take place in the interval between examinations are often of crucial importance For example a child who has eaten unripe apples complains a few hours later of cramps in the stomach When asked where the pain is most severe he points to his navel Abdominal examination is negative but on re examination three hours later he has a tenderness and rigidity in the right lower abdominal quadrant acute appendicitis is then confidently diagnosed Or a man of fifty after a city banquet has indigestion —epigastric pain radiating to the interscapular region and accompanied by belching His doctor finds nothing abnormal on examining the abdomen or chest but notices that the patient is restless and appears somewhat shocked the pulse is rapid and the systolic blood pressure low He suspects myocardial infarction An electrocardiogram taken at this time is within normal limits But when this is repeated twelve hours later although the clinical picture has improved characteristic cardiographic changes which support his earlier suspicion are found

Together the history and examination and such additional investigations as they may suggest will in most instances reveal the diagnosis in terms for example of pneumothorax gastric ulcer aortic incompetence etcetera but it must never be forgotten that for *a complete diagnosis* (and a diagnosis must be complete if all the necessary preventive and therapeutic measures are to be carried out) it is important to know *the sites of disease what functional (including mental) disturbances accompany the disease and what are its causes* For example in pneumothorax management demands that the cause—subpleural tuberculosis ruptured emphysematous bulla etcetera—must be determined in gastric ulcer whether it is simple or malignant in aortic incompetence whether it is syphilitic atheromatous rheumatic or associated with subacute bacterial endocarditis

It may be recalled that groups of symptoms which recur in similar patterns (*syndromes*) and which are often referred to as diseases are simply pointers to the site functional disturbances or causes of disease Staccato speech intention tremor and nystagmus are not the classical triad of a disease —dissemin ated sclerosis—but of cerebellar dysfunction whatever the cause Similarly tinnitus deafness and vertigo (Menière s disease) may be the expression of any cause of inner ear dysfunction Diabetes mellitus is a syndrome pointing to impaired carbohydrate toler ance and not necessarily to pancreatic disease Recurring rigors on alternate days strongly suggest as their cause benign tertian malaria

What determines *the significance of symptoms and signs in diagnosis*? Firstly the *frequency* of their presence in any disease and secondly their *specificity* for that disease [1]

For example vomiting is an almost constant accompaniment of pyloric stenosis *i e* its frequency is almost 100 per cent [2] But vomiting is a symptom of a host of other conditions *i e* its specificity for pyloric stenosis is very small L E cells are *found in about 60 per cent of patients with established systemic* lupus erythematosus (frequency=60 per cent) but these cells are very rarely found in diseases other than systemic lupus erythema tosus (specificity=95 per cent) In untreated general paresis a positive Wasserman reaction is found in the spinal fluid in 100 per cent of victims (frequency=100 per cent) but since it is found in many other varieties of active neurosyphilis its specificity is small

There are two important practical points which emerge from a knowledge of the frequency and specificity of symptoms and signs

First specificity increases with the number of symptoms and signs which constitute a pattern of disease For example in

[1] Here again disease is used not in the sense of a specific entity but as indicating a syndrome (of anatomical physiological or pathological significance) or combination of syndromes For a full discussion of this see the writer s Presidential Address to the Royal Society of Medicine (Section of Medicine) in the *Proceedings of the Royal Society of Medicine* (1955) Vol XLVIII p 155

[2] The figures quoted are derived from a personal series of cases others may differ a little but the general trends will be similar They are not quoted to imply mathematical exactitude but as rough illustrations

duodenal ulcer we know that the frequency diminishes but the specificity increases with the increasing complexity of the symp tom pattern

	Frequency	Specificity
1 Epigastric pain	90 /	Almost nil
2 two to four hours after a meal and relieved by food	65 %	About 30% (i e 1 in 3 patients with this complaint are found to have a duodenal ulcer)
3 recurring in attacks lasting weeks or months with in tervals of complete freedom from symptoms lasting weeks months or years	40	About 65
4 with a history of melaena	15	About 90%

Increasing specificity (and this is achieved by full history examination and necessary investigations) increases greatly the likelihood of accurate diagnosis

Secondly if a symptom or sign is commonly found in a large variety of diseases its absence is more significant than its presence in any specific diagnosis i e as a positive sign it is in itself of little value but as a negative sign it may assume great importance For example the absence of vomiting is strong evidence against pyloric stenosis and the absence of a positive Wasserman re action in the spinal fluid of a suspected general paretic who has received no treatment for the disease almost certainly excludes the diagnosis

The ideal which those with the penny in the slot machine type of mind seek is the pathognomic sign with 100 per cent frequency and 100 per cent specificity Hence the worship of the instrument and the laboratory But disease is so protean in

its manifestations that as yet scarcely any disease has a sign that can be recognized at all phases of its evolution as for example in the case of myocardial infarction already quoted although electrocardiographic changes have a high frequency and specificity

In many instances and the earlier in the course of disease the patient is seen the greater is the likelihood the evidence available is equivocal it is consistent with more than one diagnosis And yet the live problem is to be preferred to the dead certainty so we are often compelled to make a provisional diagnosis (though *prepared to change it if additional evidence* appears) and act upon it In this respect the scientific method must yield to the pragmatic As Sherrington wrote Science nobly declines as proof anything but complete proof but common sense pressed for time accepts and acts on acceptance

It is in coming to a provisional diagnosis that experience wisdom and judgement play so vital a role But it is possible to enunciate a few helpful guiding principles —

1 THE COMMONEST DISEASES ARE THE COMMONEST

Obstructive jaundice *may* be due to an aneurysm of the hepatic artery compressing the common bile duct or to an ascaris lumbricoides popping its head into the common bile duct but it is much more likely to be due for example to gall stones hepatitis or carcinoma of the head of the pancreas Pruritis vulvae *may* be the first and for some time the only symptom of Hodgkin s disease but commoner causes *e g* diabetes mellitus and local infections—should first be excluded But one must here reiterate *the importance of a complete examination if mass statistics are* not to overrule the findings in any individual patient For example constipation from simple causes is much commoner than cancer of the colon But in all cases of constipation in the adult especially of recent origin and progressive cancer of the colon must be excluded even if there be no overt evidence by a detailed investigation

2 THE DIAGNOSIS OF A CURABLE DISEASE WHICH IS CONSISTENT WITH THE CLINICAL FINDINGS IS, EVEN IF LESS LIKELY, TO BE PREFERRED FOR ACTION TO THAT OF A COMMONER BUT INCURABLE LESION

A striking illustration of this advice occurred in my practice a dozen years ago A distinguished architect was called in consultation over new Government buildings in Delhi He travelled there by air alighting at Cairo for a meal Shortly after his arrival at Delhi he complained of abdominal pain and distension but no diarrhoea he was found a few days later to have a high fever and a relatively slow pulse rate and on this evidence was diagnosed as typhoid fever No confirmatory tests were carried out but he was treated by chloramphenicol His fever however persisted and during the ensuing weeks his abdominal distension worsened he lost over two stones in weight became very constipated and in the eighth week of his illness developed an obstructive jaundice He was then flown back to England and came under my care The salient features on examination were his persisting pyrexia and progressive emaciation obstructive jaundice with an almost complete intestinal obstruction ascites and (palpable by dipping) an enormously enlarged liver with other massive tumours more widely spread in the abdomen These became more definable after paracentesis which yielded a clear straw coloured fluid free from tumour cells The clinical picture strongly suggested carcinoma of the colon (from which his father had died) with intestinal obstruction and metastases in liver and peritoneum But the history of a rapid onset with early fever after a meal in Cairo suggested the remote possibility of amoebomata Eight consecutive stools were examined by colleagues in the School of Tropical Medicine but no evidence of amoebiasis was found Nevertheless as carcinoma of the colon with metastases—almost certainly the diagnosis on the evidence—is fatal the patient was treated with emetine He is alive well and active twelve years later!

Pertinent examples of this general rule are also found in malignant hypertension if the remote possibilities of phaeo

chromocytoma or *unilateral chronic pyelonephritis* are fully investigated

3 NEVER MASK SYMPTOMS WHICH MIGHT HELP *TO ESTABLISH A DIAGNOSIS*

It is tempting especially when pain is severe at the onset of an illness to give a strong analgesic particularly an opiate This may well postpone the diagnosis until an advanced and danger ous stage of an illness has been reached as for example in acute appendicitis where the onset of generalized peritonitis may be so masked

4 DO NOT DELAY NECESSARY INVESTIGATIONS, EVEN SURGICAL EXPLORATION, TO ESTABLISH A DIAGNOSIS

(*a*) if such procedures increase the likelihood of a firm diag nosis with a better chance of survival or of preventing or reducing permanent damage and (*b*) if the hazards of these procedures are less than those of the disease which is suspected

For example if symptoms point to an early cancer of the stomach but gastroscopy and X rays are negative exploratory laparotomy is a justifiable procedure Again even when ascites or a pleural effusion is not of itself giving rise to symptoms para centesis is justified as a diagnostic measure for its risks are so small compared with the value of the information it may yield

The two provisos are well illustrated by a recent example In a man aged sixty the differential diagnosis of an enlarged liver lay between cirrhosis tuberculosis and amyloidosis The history examination and various diagnostic procedures had failed to differentiate these and the question of liver biopsy arose but it was decided against on the grounds (i) that it carried a significant hazard (ii) if the liver was tuberculous the puncture might not penetrate a discernible tuberculous focus and (iii) that of the three possible diagnoses only that of tuberculosis held out any reasonable hope of a cure or of specific treatment being given

5 DIAGNOSES SHOULD NOT BE MULTIPLIED BEYOND NECESSITY

The medical equivalent of Occam s razor viz that *diagnoses should not be multiplied beyond necessity* must always be heeded

A single diagnosis which covers the available evidence is to be preferred to multiple diagnoses For example if a patient with cancer of the breast or a history of a radical mastectomy develops signs of an intracranial tumour the pathology of this should be metastatic breast cancer If a diabetic or alcoholic develops a peripheral neuropathy it would be superfluous unless there were special pointing features to look for porphyria But it must not be forgotten that more than one disease *may* occur in the same patient For example not every disease occurring in a patient who has had syphilis and still has a positive Wasserman reaction is of syphilitic origin

It is perhaps well here to emphasize that *if a symptom or sign cannot be fitted into a single diagnosis it must not be forced into it* Nor must symptoms or signs however trivial they might appear which fail to fit into a diagnosis be deliberately ignored As Agatha Christie s famous detective Hercule Poirot declared Beware! peril to the detective who says It is so small that it does not matter It will not agree I will forget it That way lies confusion—everything matters The case of amoebiasis described earlier illustrates this point It was the sudden onset with early pyrexia which raised the possibility of amoebiasis in a patient whose signs were overwhelmingly in favour of a diagnosis of colonic cancer

6 NEVER FORGET THAT THE DIAGNOSIS OF 'FUNCTIONAL DISEASE' IN THE SENSE OF 'PSYCHOGENIC' IS THE COMMONEST REFUGE OF THE DIAGNOSTICALLY DESTITUTE

This must never be made solely on the absence of signs of organic disease There must be positive signs before a psychogenic illness is diagnosed and many a physician has rued

the day that he overlooked the fact that even the long established neurotic patient may develop organic disease

7 NEVER ALLOW THE INITIAL PROVISIONAL DIAGNOSIS TO BIAS LATER OBSERVATION AND JUDGEMENT AS THE COURSE OF AN ILLNESS UNFOLDS ITSELF

The rapid onset of a hemiparesis in a man of fifty with arteriosclerosis and hypertension is justifiably regarded as a cerebral vascular lesion until increasing paresis papilloedema stupor etcetera point to the possibility of a cerebral tumour Again in the patient with a gastric ulcer diagnosed as simple because on treatment with alkalis the pain disappears appetite improves and weight is gained for several weeks or even months but then regresses the prejudice of the initial diagnosis should not blind the doctor to the possibility of cancer when the improvement is halted despite treatment

When one has looked after the patient often for many weeks or months it may be difficult to take a wholly objective view This is the time for a consultation

8 AMONGST THE MOST SIGNIFICANT OF SYMPTOMS ARE THOSE WHICH RELATE TO A CHANGE IN NORMAL HABITS

When a man of fifty who has a daily bowel action finds that he has to take increasing doses of aperients the possibility of an increasing intestinal obstruction is much more likely than in a man who has suffered from chronic constipation for years Similarly a man of forty five who finds himself breathless on walking up a flight of stairs which previously caused no dis comfort or who has always enjoyed food and finds that his appetite is failing will almost certainly be in the early stages of organic disease And finally—

9 NEVER ALLOW THE SOCIAL POSITION OF A PATIENT TO LIMIT YOUR EXAMINATION

Cancer of the rectum for example is no respecter of persons and the old adage— If you don t put your finger in it you put your foot in it —is equally applicable to rich and poor to commoner and nobleman Nor should rank blind one to the possibility of such diseases as syphilis for every bishop has been an undergraduate and every admiral a midshipman

The aim of early diagnosis as we have earlier expressed is to discern an illness early in its course for it is then that its control is best achieved But despite the emphasis on early diagnosis the fact must be frankly faced that often the patient first presents when his disease is well advanced sometimes because he has neglected early symptoms (and only public education in personal health will help to prevent this) but also because the early symptoms may not occur until the disease is advanced and incurable For example haemoptysis may be the first sign of a cancer of the bronchus already extensive and inoperable and a husky voice or a spontaneous fracture the first evidence of cancer of the thyroid

The qualities demanded of the diagnostician are both *humane* —sympathy kindness toleration and consideration and *scientific*—the ability to observe accurately and meticulously and to draw the correct inferences from these observations It cannot be too strongly emphasized that for the latter clinical experience is not enough The atypical diagnostic problem will be solved only by those who are well grounded in human anatomy physiology and pathology—the firm tripod on which the science of medicine is founded

CHAPTER II

THE ACUTE ABDOMEN

BY A G R LOWDON

THE well worn term the acute abdomen covers a wide variety of conditions most of which call for emergency operation but the subject of this chapter must include a consideration of various diseases which may present with acute abdominal symptoms and which in practice must be differentiated from conditions requiring operation

The importance of early diagnosis in acute abdominal disease requires no emphasis One feature common to all these conditions is that they are liable to lead to complications which involve serious morbidity and perhaps a fatal outcome unless treatment in most cases surgical treatment is instituted at an early stage The diagnosis in any patient with acute abdominal symptoms is therefore approached with a sense of urgency by general practitioner and surgeon alike

Both the practitioner and the surgeon must strive from first to last to make a precise and accurate diagnosis of the disease affecting the patient but it must also be admitted that the important practical decision for the practitioner is the answer to the question must this patient go to hospital? and the critical question for the surgeon is should this patient have an operation and if so what is the best abdominal incision to use? In other words the best diagnostician must remember that he may be wrong for he certainly will be wrong sometimes having made his diagnosis sometimes in terms of certainty sometimes in terms of probability he must answer the more important question what then is the treatment which is safest and best for the patient? Diagnosis is only a means to this end

There is a danger unfortunately that the evident importance of the practical decisions—for or against transfer to hospital

and for or against operation—may lead to diagnostic laziness Accurate diagnosis must be regarded not as an academic exercise but as the best possible way of arriving at the right treatment

THE CLINICAL HISTORY

It may be a platitude to say that early and accurate diagnosis depends above all on detailed study of the clinical history of the patient but it must be said again and with special emphasis in relation to the diagnosis of abdominal emergencies The physical examination of a patient may be interpreted quite correctly as showing evidence of peritonitis or intestinal obstruction but it will seldom permit a reliable diagnosis of the *cause* of the peritonitis or the intestinal obstruction Findings on examination may even be seriously misleading unless the history has been taken carefully An irreducible hernia may assume undue significance in the mind of a doctor who has not discovered that it has been irreducible for two years and that the abdominal pain has been constant and not colicky in character Pain rigidity and tenderness in the right side of the abdomen may be readily misinterpreted if the doctor has failed to elicit the history that the pain is felt only on inspiration and coughing

PAIN

The careful analysis which should be made of any symptom can be well illustrated with reference to abdominal pain which is the presenting complaint in most cases of acute abdominal disease

The **situation** of the pain should be learned not only from the patient s description but also by asking him to demonstrate by pointing where the pain is felt Valuable information may be gained by noting how the patient indicates the site of the pain —with one finger on a precise spot or with a hand moving vaguely over the abdomen To learn that a colicky pain is felt in a vaguely central situation is not enough It may be epigastric suggesting an origin in the foregut umbilical suggesting an origin in the midgut or definitely in the lower abdomen as is common with pain arising from the distal colon By contrast with the

indefinite localization of intestinal colic the pain of renal colic will be more clearly located on one or other side and the situation of other pains like those of acute duodenal ulcer or appendicitis may be indicated by the patient pointing with one finger

The **severity** of the pain must be assessed carefully The patient s answer to the question was it a severe pain? is not enough It is more informative to ask the patient how he or she behaved when the pain was severe A patient may walk about the room restlessly with the pain of peptic ulcer but he is unlikely to do so if a perforation has occurred A renal or biliary colic will usually make the patient writhe or want to cry out in agony The pain of diverticulitis for example may be described as severe but will not make the patient behave in this way A parous woman can help by comparing the pain to those of childbirth renal and biliary colic will usually be described as being much worse than labour pains

The **onset** of the pain should be described in detail and any statement that the onset was sudden should be confirmed by asking the patient what he or she was doing when the pain started The very moment of onset of the pain in perforated peptic ulcer can be remembered in this way and the colics may have an equally dramatic start but the pain of inflammatory lesions is unlikely to be timed so precisely Information about the **progress** of the pain since its onset is often helpful it may have got steadily worse as may be expected of the pain in a progressive inflammatory lesion or it may have settled after a period of great severity to be replaced by a dull ache as is often the case after renal or biliary colic The abatement of the pain of perforated ulcer after a few hours when exudate begins to dilute the irritant gut content is well known as the period of illusion and is not to be mistaken for improvement in the patient s condition The duration of a transient pain or the periodicity of an intermittent pain as in intussusception must also be noted

The patient may be asked to describe the **character** of the pain Some terms such as dull ache like toothache or coming and going' may be helpful but other common descriptions such as sharp or knife like are less so In this connection it is helpful to ask the patient to compare or contrast the pain with

that of previous experiences which the patient can remember as for example labour pains or those of any previous abdominal disease of which the diagnosis is known

Change of situation of the pain may be of great importance The change in location of pain in typical appendicitis—first in the upper abdomen and then in the right lower abdomen—is well known but pain from other causes may change its location in a similar way The first pain from the small perforation of a duodenal ulcer may be felt in the right upper abdomen with subsequent shift to the right lower abdomen when intestinal content trickles over the transverse colon and omentum

The **radiation** of some types of pain is typical and should be distinguished from a history of change in location The radiation of biliary pain round the lower chest or of renal colic to the groin or genitalia is particularly helpful Sometimes the description of pain in other situations is revealing and as these secondary pains may be eclipsed by severe abdominal pain the doctor should remember to ask for this kind of information Shoulder tip pain may provide valuable evidence of subdiaphragmatic irritation in patients with cholecystitis perforated ulcer or ruptured spleen this pain of phrenic reference may occur of course in any patient with generalized peritoneal irritation even if the origin is in the pelvis as in the case of ruptured ectopic gestation Another rare but interesting distal pain is that felt on the medial side of the thigh and knee from irritation of the obturator nerve by an obturator hernia

Finally the history must include description of **previous attacks** of abdominal pain whether similar or different Common examples of the importance of the previous history are found in the account of hunger pain usually but not always given by a patient who has a perforated ulcer and in the recurring lower abdominal colic which often precedes intestinal obstruction by a scirrhous carcinoma of the distal colon

VOMITING

Other symptoms must be analysed with the same attention to detail Of these vomiting is nearly as common as pain It is not enough to know that the patient has vomited We must learn

when the vomiting started and its relation to other symptoms whether it was an isolated occurrence repeated a few times or many times the character and if repeated the changing character of the vomitus may be significant

The significance of obvious blood in the vomit or of repeated and finally faeculent vomiting is not likely to be overlooked but more subtle features may be revealing For example the symptoms and signs of two upper abdominal crises perforation of peptic ulcer and acute pancreatitis may be very similar and easily confused but sometimes a history of repeated vomiting and retching which is rare in perforation may be the one feature of the illness suggesting that the correct diagnosis is pancreatitis Various other conditions may be associated with persistent and severe vomiting—as in renal or biliary colic or when an ovarian cyst has twisted its pedicle—but it is only when a mechanical obstruction or peritonitis causing paralytic ileus affects the bowel that the character of the vomitus gradually changes from gastric and then bile stained to the brown offensive material which comes up from the small intestine

Some patients naturally vomit readily others vomit rarely and only when they have some fairly serious abdominal upset It is sometimes worthwhile to find out to which of these groups the patient belongs mere loss of appetite and nausea in the latter group may have more significance than an episode of actual vomiting in the former group

OTHER SYMPTOMS

Other symptoms of abdominal disease may be less common than pain and vomiting but they are no less important

Any history of recent diarrhoea or constipation must be noted In this connection two traps for the unwary deserve mention A history of several loose motions which may be described as diarrhoea may be given by a patient who has appendicitis this occurs more often in children and particularly if the inflamed appendix is lying in the pelvis It must be remembered also that diarrhoea may occur with a strangulated hernia of Richter s type and the association of diarrhoea with other findings which suggest

intestinal obstruction or strangulated hernia should bring this possibility to mind

Enquiry about the bowel function should be accompanied by enquiry about the taking of laxatives Recent diarrhoea may be the result of ill advised—and undeclared—purgation The chronic constipation which precedes acute obstruction from a string stric ture of the colon may be disguised by steadily increasing doses of laxative medicine the patient may deny constipation but will admit the increased used of purgatives

A history of abdominal distension may be very significant, but should be accepted most critically and given weight only if the patient can confirm his impression with circumstantial detail—such as the finding that clothes have had to be loosened

Disturbances of micturition may be helpful but also mislead ing It must be remembered that inflammatory lesions such as pelvic appendicitis may irritate the bladder producing some frequency and even occasionally dysuria Moreover an inflamed appendix lying close to ureter or bladder may be the cause of a slight excess of pus cells in the urine

In a woman the menstrual history must never be neglected and it is not enough to know simply that the menses are regular The diagnosis of tubal gestation may be revealed only by the finding that the last menstrual loss was less than usual and the ruptured ectopic may occur with uterine bleeding at the expected time but of unusual character

Finally no possible clue in the past history should be over looked the vital information needed for accurate diagnosis may be found in a history of previous dyspepsia previous severe illness or previous operation

THE CLINICAL EXAMINATION

The best indeed the only way to attack the problem of diagnosis of an acute abdominal complaint is to formulate a differential diagnosis from the history alone to narrow this down still further by asking additional questions each for a specific reason (but never of course phrased in leading or suggestive terms) and then to use the examination of the patient simply

to confirm or exclude the alternative diagnoses that have been so formulated The stage of examination should be reached with a short list ready for final selection.

There is a real danger that the hurry of modern life and modern medicine will tempt the doctor to skip the details of the history which especially from a patient who is talkative and not intelligent may take so much time There is an urge to get down to the business of palpating the abdomen and getting the matter settled it must be resisted

If the examination is to take its proper secondary place in the business of diagnosis its limitations as a source of information must be remembered Localized tenderness and muscle resistance in the right lower quadrant of the abdomen maximal at McBurney s point do not point to any one disease Even when distant causes such as right basal pleurisy are excluded the finding means no more than that peritoneal irritation is present in this abdominal area There are many acute abdominal conditions in which this finding may present and no interpretation of the finding other than 'local peritoneal irritation is justifiable except on the basis of the history and other signs Nor does a positive Murphy s sign necessarily mean that a patient has cholecystitis

At the risk of labouring a simple point it may be expressed in a different way If the evidence of the history and the physical examination appear to disagree the history is often the more reliable When a patient gives a typical history of perforation of a duodenal ulcer the finding that tenderness is maximal at McBurney s point need not disturb that diagnosis when a patient gives a typical history of acute appendicitis the absence of tenderness and resistance in the right lower quadrant will not exclude that diagnosis

GENERAL CONDITION

The examination of the patient starts with an assessment of the general features which will show how much systemic upset is present Most acute abdominal emergencies should be diagnosed before these signs are severe but the finding of a patient with high fever evident toxaemia or dehydration will heighten the

sense of urgency in the doctor to make a diagnosis and begin treatment

In the general examination of a patient complaining of abdominal pain the attitude and behaviour of the patient should be noted A patient with colic is often restless or at least not unwilling to move in bed while a patient with an irritating peritoneal exudate from perforation of an ulcer is usually unable to change his position without aggravation of pain and prefers to lie immobile

The condition of the tongue is not a reliable guide except when its dryness is evidence either of severe dehydration or of uraemia

The patient s temperature pulse and respiration rate will be noted remembering that any notable change in these in the early stages of an illness should lead to the more careful exclusion of conditions outside the abdomen such as respiratory infections or coronary thrombosis High temperatures especially if preceded by a rigor should direct attention also to the urinary tract

ABDOMINAL EXAMINATION

The abdominal examination should follow the time honoured routine—inspection palpation percussion and auscultation and to these may be added examination for cutaneous hyperaesthesia In addition any doctor who examines an acute abdomen should have two obligations clearly in his mind—first to inspect all the hernial orifices and secondly to make a digital rectal examination

On inspection of the abdomen it is particularly important not to miss any localized swelling or the sign of visible peristalsis

On palpation (with warm hands) the first examination should be very gentle and should start far from the place where trouble is expected Muscle rigidity should be tested if possible before tenderness has been elicited The significance of symmetrical resistance is always doubtful but persistent asymmetrical tightness of the muscle must be noted Tenderness is one of the findings on which the doctor is most dependent In acute abdominal disease it usually means peritoneal irritation but as

has been emphasized already other information is required to elucidate the cause of this Similarly a mass is only a mass—not a diagnosis A tender mass in the right iliac fossa is often an appendix abscess but it is sometimes an abscess related to a carcinoma of the caecum sometimes the thickened bowel of regional ileitis or ileo caecal tuberculosis and sometimes a red herring —like an ectopic kidney

Percussion is useful in the detection of free peritoneal fluid the distended bladder etc There is a current fashion to belittle the value of percussion of the liver dullness in an attempt to reveal the presence of free gas in the peritoneal cavity With this the present writer cannot agree If the doctor remembers the limitations of the sign—that liver dullness is reduced when the liver is shrunken by cirrhosis rotated upwards by distended bowel or partly covered by emphysematous lung—he will find it as reliable a sign as many others Definite loss of liver dullness especially if noted in the mid axillary line is often a helpful finding strongly suggestive of escape of gas from perforated bowel

Auscultation of the abdomen is another valuable source of information—if the observer has trained himself by experience to recognize the normal Excessive peristaltic activity may be helpful in permitting the early diagnosis of a mechanical obstruction even before distension or repeated vomiting appear A completely silent abdomen strongly suggests the inhibition of all peristaltic movement by peritonitis But no rule is true of all cases normal bowel sounds may be present with peritonitis and the excessive activity of the earlier stages of mechanical obstruction may be followed by paralytic ileus and silence in the later stages

Last but certainly not least comes the rectal examination Pelvic appendicitis may be missed with disastrous consequences if this examination is omitted It may be less serious but it is *no less shameful to miss the occasional case of faecal impaction* in the rectum which may present either with spurious diarrhoea or with obstructive symptoms

ADDITIONAL INVESTIGATIONS

The only additional investigation normally available to the general practitioner is the examination of the urine This can be tested for albumin with equipment carried in the doctor s bag but if the patient should be seen in the consulting room it is more informative—and easier—to put a drop of uncentrifuged urine on a slide under the microscope and look for abnormal cells

In hospital it may be helpful to make a white cell count a confirmed low count is often more helpful than the more usual finding of a moderate leucocytosis In acute appendicitis for example some leucocytosis is invariably present by the time that the patient is showing systemic upset and the finding of a low white count in a patient with abdominal pain and fever is against the diagnosis of any pyogenic inflammatory lesion

Plain X ray examination of the abdomen may be carried out in hospital and sometimes provides valuable information X rays are taken with the patient in both recumbent and erect positions Particularly significant findings are the presence of free intra peritoneal gas under the diaphragm showing that there has been an escape of gas from the bowel or the picture of abnormally distended coils of bowel indicating that there is an obstruction of the intestine Interpretation of the gas patterns in a case of obstruction may permit a fairly accurate inference about the level of the obstruction or even sometimes a precise diagnosis such as that of volvulus of the colon

Another valuable special investigation is the estimation of serum or urinary diastase which sometimes permits the confident diagnosis of acute pancreatitis in an otherwise doubtful case

In the last resort it may be safer to look and see than to wait and see The abdominal surgeon frequently has to be content with a decision to perform laparotomy on the basis of a provisional diagnosis or indeed on a diagnosis as indefinite as peritonitis possibly due to X or intestinal obstruction cause unknown

APPENDICITIS

Recent advances in surgical management have resulted in a steady fall in the mortality of acute appendicitis but it is still a dangerous disease and the first essential in the struggle against it is still early diagnosis It is clearly impracticable for the general practitioner to send to hospital as suspect every patient with pain in the right iliac fossa there are not enough hospital beds The selection of cases must depend on judgement based on a great many factors no categorical advice or statement would be safe but a few general remarks may be helpful

If the doctor sees the patient in the very early stages when pain is still central the diagnosis of appendicitis will be justified only if local signs in the iliac region are definite otherwise the patient should be seen again after a few hours

When the symptoms and signs of right lower abdominal peritoneal irritation have been preceded by definite central abdominal pain nausea and vomiting these should be taken as a warning that the appendicitis if such it is is of obstructive and more dangerous type calling for prompt action When *the pain seems to start in the right lower abdomen there is more* difficulty If the local physical signs are definite and consistent there is no excuse for delay but if the signs are equivocal and more particularly if the patient has often before had pain in the right iliac fossa it may be right to wait for the development of fever or more definite local signs before sending him to hospital

It should be remembered that localization of peritonitis after perforation of an inflamed appendix is less efficient in children than in adults It is correspondingly more important in the case of a child to make the diagnosis and undertake operation at an early stage

Some of the serious difficulties in the early diagnosis of acute appendicitis arise from the very variable situation of the appendix Pelvic appendicitis may be so insidious in its onset that the patient does not consult the doctor until perforation has occurred and even if the doctor is consulted early the symptoms and signs are vague and inconclusive—until a rectal examination is made Other difficulties arise when an inflamed high or sub

hepatic appendix tends to be confused with upper abdominal conditions such as perforated duodenal ulcer or acute chole cystitis The retro ileal appendix may be the cause of repeated vomiting with poorly localized physical signs

PERFORATED PEPTIC ULCER

The typical case of perforated ulcer is so dramatic and so severe that delay in diagnosis is most unlikely to occur but when the perforation is small and only a small leak occurs the diag nosis may be more difficult The leaking fluid may trickle down over the front of the transverse colon and omentum and the patient may present with localized symptoms and signs either in the upper or in the lower abdomen It is in these cases that the finding of loss of liver dullness may be very helpful because not infrequently more gas than fluid escapes through a small perfora tion An X ray for pneumoperitoneum is of course more reliable However free gas under the diaphragm is not invariably present with perforation and its absence does not exclude the diagnosis

In the typical case with generalized abdominal pain and boarded muscles the patient is usually found lying flat on his back and frightened to move It must however be realized that other attitudes are compatible with the diagnosis sometimes the patient sits up and will not lie down even to be carried on a stretcher and rarely the patient is found throwing himself about in a hysterical manner

INTESTINAL OBSTRUCTION

When the symptoms and signs of intestinal obstruction are fully developed the diagnosis presents no difficulty—but by that time the patient is exhausted dehydrated with reduced blood volume and possibly also with peritonitis if the obstruction is of a strangulating type The aim must be to diagnose intestinal obstruction before the clinical picture is complete The earliest features are colicky pain vomiting and increased intestinal sounds on abdominal auscultation If there is an incarcerated hernia the diagnosis is easy In other cases it may be possible

even at this early stage to feel by careful palpation one or two distended coils of intestine or failing this help in diagnosis it may be possible (in hospital) to obtain confirmation of this diagnosis by finding abnormally distended coils of bowel in a plain X ray

At a later stage there may be constipation and abdominal distension but it is not necessary or desirable to wait for evidence of these Faeculent vomiting and the signs of peritonitis indicate that the disease is far advanced

In high small intestine obstruction vomiting will be severe and distension minimal or absent On the other hand when the obstruction is in the distal colon often due to scirrhous carcinoma of the colon the patient may be accustomed to occasional low abdominal colic and may ignore this and one early reflex vomit He reports to his doctor only when he finds he is unable to pass flatus and the abdomen is becoming distended In these cases the diagnosis should be made at this stage and before repeated vomiting supervenes

Diagnosis is an urgent matter in all cases of intestinal obstruction but particularly when there is any risk of strangulation of bowel This risk is always present with incarcerated hernia while in other cases where the cause of the obstruction is intra abdominal and obscure strangulation may be suggested by the finding of local abdominal tenderness and the relatively early development of fever and other signs of systemic upset

Paralytic obstruction of the intestine may present without any colic and with absent bowel sounds but it is usually a complication either of neglected peritonitis or of operation

INTUSSUSCEPTION

Intussusception the common cause of intestinal obstruction in infants deserves special mention The mortality of this disease occurs entirely in the babies who are found at operation to have irreducible or gangrenous intussusception because of late diagnosis Again it must be emphasized that some of the 'classical signs of the condition are late signs The diagnosis should be made before there is much abdominal distension and before a blood stained stool is passed certainly before the intussuscep-

tion can be felt on rectal examination! In other words it should be our aim to diagnose the condition on the typical history from the mother that the baby has had attacks of screaming with pain when he draws up his knees and looks pale There may have been some vomiting If a typical tumour can be felt at this stage the diagnosis is certain but palpation of the tumour in an unco operative child may have to wait until the child is admitted to hospital and given a sedative

OTHER ACUTE ABDOMINAL CONDITIONS

Many other conditions may be the cause of acute abdominal symptoms but this is not the place for a complete or detailed review Of these **acute cholecystitis** is perhaps the most common Since this condition usually settles with conservative treatment there may not be the same urgency about early diagnosis but it is important to recognize the case which is not improving and to anticipate perforation of the gall bladder by hospitalization and operation **Acute pancreatitis** may be difficult to distinguish from perforated ulcer or other causes of severe upper abdominal pain but it usually presents with unequivocal signs of periton itis and there is little difficulty in deciding that the patient should be sent to hospital where diastase estimations may help

The various forms of **colic** may cause extremely severe abdominal pain Renal and biliary colic usually have fairly characteristic features and very characteristic severity they only rarely call for urgent surgical treatment Intestinal colic has been considered in its relation to intestinal obstruction but it must be remembered that severe intestinal colic may occur in other diseases—for example in enteritis lead poisoning or con stipation

Among the rarer causes of severe abdominal pain which must be kept in mind are **dissecting aneurysm** of the aorta and rup tured aneurysm of the abdominal aorta

Of the gynaecological conditions which give rise to acute abdominal signs **salpingitis** and complications of small or large **ovarian cysts** are the most common Among the more serious is the **ruptured ectopic gestation**, the diagnosis of which may rest

largely on a careful analysis of the menstrual history It should of course be the doctor s aim to diagnose ectopic gestation before rupture occurs

Finally it is important to remember the extra abdominal conditions which may cause abdominal pain, such as **coronary thrombosis, diaphragmatic pleurisy, spinal disease,** and **tabes dorsalis** the tabetic crisis is more likely to be forgotten as it becomes rarer **Bornholm disease, porphyria,** and **acute rheumatism** beginning with abdominal pain simulating appendicitis are also notorious traps for the unwary

CHAPTER III

THE ACUTE SPECIFIC FEVERS

By G I Watson

FOR the space of fifteen years from 1661 to 1676 Thomas Sydenham studied Epidemick Diseases in London and about them he was able to say Tho these Diseases may seem alike to the Unwary because in some sort they do agree to out ward Appearance yet if you seriously consider they are very different To guide his consideration of them he had only his unaided senses so it continued until the microscope began to reveal other minute differences between one patient s illness and another s We can now classify infectious diseases into three broad groups The first consists of those such as chickenpox measles and mumps which are clinically recognizable because of changes in the body which are visible to the naked eye In the second group—for example influenza undulant fever or virus meningitis—some of the diagnostic signs are microscopic or serological and cannot be elicited at the bedside by the doctor unaided The third group comprises a large but diminishing remainder of short fevers as yet poorly differentiated at the bed side and in the diagnosis of which no laboratory assistance is yet possible In this chapter I shall try to show how infections in the first two categories can be suspected or diagnosed early—sometimes before the clinical picture has fully developed—realizing however that in general practice in this country the short unnamed fevers are still the most numerous group

By definition these patients are feverish They are usually children the onset of their illness is often acute and the cause of it is an identifiable infective agent—either bacterium or virus Some infectious fevers are commoner than others even those which are frequently seen vary in prevalence not only from one season to another but in different continents and countries It

is undoubtedly easier for a doctor to make a correct early diag nosis if the disease from which his patient is suffering is one which he has borne in mind in this country a sporadic case of malaria is more likely to be missed than it would be in the tropics while a correct early diagnosis of measles before the rash develops is more likely to be made in a known contact than in the first patient of an epidemic The purpose of this chapter is to help a doctor as he considers the outward Appearance' of a feverish patient and to appreciate some of the traps for the Unwary

PRACTICAL IMPORTANCE OF EARLY DIAGNOSIS

Among several reasons which finally lead the parents of a sick child to ask for help is their anxiety lest their gravest fears be well founded Tell me the worst doctor is it polio? His breathing was so fast we were afraid to go through the night. First in importance when making an early diagnosis the writer would put the restoration of parental peace of mind Disturbed nights and extra work by day are more easily borne when one s strength is not being leeched by anxiety Even when the doctor is still medically uncertain about the true nature of an infection something about the case may enable him to set their worst fears at rest If a doctor has unfortunately at times to confirm an un welcome diagnosis such as meningitis the knowledge that their child s danger has been recognized in good time often appears to give strength to those who must wait upon the outcome of its treatment

One of the first things to do with a new doll or a new disease is to give it a name After a good night s sleep a sick child wants to know what he has got There is something comfortably familiar about the words measles chickenpox and nettle rash and something unsatisfactory about the yet unnamed infections caused by strains of adenovirus or the Echo or Cox sackie viruses Virus pneumonia and virus meningitis con jure up too alarming a picture in the mind to be comfortable as names for these usually self limiting and relatively benign infec tions One such outbreak in my practice due to Coxsackie virus

A patient s next concern is usually with the duration of his illness How long am I going to be here? For an ill mother there is a question of arranging with someone to run her house the man of the house wants to know how long it will be before he returns to work and a child s hope will centre on being up in time for a birthday or some other treat in store Where the duration of high fever may often be accurately forecast as in measles or influenza this information should be given at the first or second visit and in influenza where the possibility of a second bout of high fever exists it is reassuring for the relatives to be told that such a rise in temperature would not necessarily mean that complications had set in In winter vomiting disease someone else is often affected two nights after the onset of the patient s symptoms a warning that this may occur helps both the parents arrangements and the doctor's reputation The knowledge about his patient s temperament which a family doctor comes to acquire helps him to decide when to tell the patient or his relatives that an illness such as glandular fever is going to last a few weeks or months rather than a few days The sooner the diagnosis is made the sooner may this problem be faced instead of having it raised inopportunely

Apart from questions about the patient s own condition others will be asked about the likelihood of illness occurring among close contacts about the probable date of onset of this and the dates when contacts will in turn be infectious to their friends These matters become particularly acute at the end of school holidays when both the doctor and his patients are conscious of their duty to prevent the introduction of infectious illness into boarding schools A problem which a boarding school doctor sometimes has to face when making a tentative early diagnosis of some potentially serious disease is whether the school should therefore be closed In all problems of this sort the family doctor will be wise if he establishes early in his career the friendliest contact with the Medical Officer of Health of his local authority It is to him that the practitioner should feel able to turn if in doubt for help and advice about the diagnosis and management of infectious diseases

Finally the need should be stressed for collecting diagnostic material such as blood swabs or faeces for examination in a laboratory In an increasing number of diseases special haemato logical and virological tests are among the only means whereby a final diagnosis can be established Sometimes though certainly not always such diagnostic material can only usefully be collected during the first one or two days of fever for example the isola tion of the Asian strain of *influenza virus A* appeared to be less uncertain during the stage of rising fever than after the peak of the temperature had been reached Anyone who has attempted this type of work in general practice knows how easy it is not to have the proper facilities for collecting and storing the necessary material at the time when the patient is first visited

THE FIRST SUSPICIONS

When a child takes ill with an acute specific fever its mother is usually the first person to notice that something is wrong This she discovers on one of the several occasions when she helps or handles her child Since fever produces its most troublesome and obvious effects when at its height usually in the evening it follows that a doctor is often first summoned to such patients between six o clock and ten o clock at night

Some signs and symptoms associated with illness may force themselves strikingly upon a mother s attention at other times of the day—a rash when dressing her child or after a bath has made it more pronounced a headache for which he has been sent home early from school by his teacher a convulsion in the evening when she has got the living room well heated before putting the baby to bed vomiting in the evening after supper or when he has been in bed for a short while and of course the sleeplessness of a coughing or crying child which also gives her husband and herself a bad night On the other hand she is apt to overlook as commonplace the sneezing of measles and the sore throat of scarlet fever when these are present in the morning before school Indeed it is quite remarkable how many ill children are wrongly thought to be lead swinging in the morning only to be sent off to school feeling miserable infecting

their playmates but learning nothing themselves before being sent home early by an observant teacher Swellings however are often noticed at breakfast time and I seldom get a first call for mumps late in the day

Unlike the mother who has to make a quick decision perhaps on a cold wintry morning between attending to her other chores the schoolmaster has longer to make up his mind about a pupil who does not seem well At school headache is by far the commonest symptom complained of by a child the teacher s attention is more likely to be attracted if the child is coughing or has vomited

By the time therefore the doctor comes on the scene two three or sometimes more people have already decided that little Billy or Mary is not well—the child its parent its teacher and—as often as not—grannie too The burning question is What is it? Measles? Chickenpox? Flu? — Diphtheria? (not often now thank goodness) But therein lies a danger for any doctor who eventually finds himself faced perhaps only once in his career with an unfamiliar but serious and killing disease which was commonplace to his father or grandfather He may well ask whether it is reasonable even to expect him to bear diphtheria or smallpox in mind as an alternative diagnosis in Great Britain at the present time Yet in 1956 eight children died of diphtheria out of fifty one notified cases and from time to time smallpox reappears unexpectedly

It cannot be stressed too strongly that any appearance of exudate or thrush on the palate or uvula or perhaps on one tonsil should awaken a suspicion that the child might conceivably have diphtheria If the habit of throat swabbing has not been allowed to atrophy (as alas! it is in danger of doing with the arrival of wide spectrum antibiotics) then the bleeding caused by disturbing a diphtheritic membrane will increase any previous doubts about the correct diagnosis Follicular pus and the confluent exudate seen in staphylococcal infection do not leave a bleeding surface when removed There is a putrid smell about the breath in both diphtheria and Vincent s angina which may attract attention even when they are encountered for the first time

The early diagnosis of smallpox from chickenpox again depends upon one s suspicions being aroused at the first visit. A child starting chickenpox is seldom so ill as to want to stay in bed when the first spots on the trunk are noticed only when the second or third crop of spots comes out, the temperature rising with each crop does he begin to feel miserable with the irritation and fever It is rare to see chickenpox blisters on the hands or feet during the first or second crop Early toxaemia and a scanty rash on the back and abdomen should certainly make one look more closely at the form of the blisters them selves and should make one telephone the Medical Officer of Health A system of two way notification is undertaken by a few local authorities whereby each practitioner informs the Medical Officer of Health from time to time about fresh out breaks of infectious illness as they occur whether these are statutorily notifiable or not the local authority in turn sends out a weekly summary of such information obtained through the doctors and schools to all general practitioners in its district By this means neighbouring doctors are reminded about prevalent *infectious diseases which may not yet have affected their own* patients

INITIAL SYMPTOMS AND THEIR DIAGNOSTIC VALUE

At the start of many feverish illnesses one or more of the following symptoms have usually been noticed by the patient or those about him before medical advice is sought headache lassitude photophobia vomiting or diarrhoea sore throat crying irritability loss of appetite coughing or sneezing aching in the back or limbs Something will now be said about the diagnostic value of each of these symptoms during the early stages of specific infections

While mild **headache** is common in almost any feverish illness as a presenting symptom it is nearly always of diagnostic import ance During the 1957 epidemic of Asian influenza in Britain headache was commoner than any other symptom during the first twelve or twenty four hours of illness it then usually passed off before the fever In acute bacterial meningitis however

headache is persistent usually occipital rather than frontal and is scarcely relieved by ordinary analgesics Headache is also severe in smallpox unlike chickenpox in typhoid and infective hepatitis it is a common presenting symptom

Lassitude is a feature noticed by or about a patient whose illness has begun insidiously as in undulant fever or tuberculosis rather than dramatically as in Bornholm disease Further enquiry may also reveal that the invalid has been taking an antipyretic to enable him to keep about in spite of his infection

Any degree of **photophobia** in a feverish patient must raise the suspicion that he is already suffering from some type of meningitis particularly since it is not as a rule one of the earliest symptoms to appear in this disease Mild degrees of discomfort in bright light may however be present in many virus diseases where the conjunctivae are slightly reddened as by measles and influenza Curiously however it is not a feature of pharyngo conjunctival fever due to adenovirus infection in which the eyes are much redder but the discharge is not purulent Pain on moving the eyes may be present in Bornholm disease

Vomiting as a presenting symptom of infection varies in frequency with age and in character with the type of infection It always calls for careful enquiry about abdominal pain and diarrhoea and also about the recent consumption of insoluble aspirin or other antipyretic tablets The younger the child the more likely he is to vomit once or perhaps twice at the start of fever particularly with tonsillitis but this does not continue Initial vomiting is rare in diphtheria more common in scarlet fever A single vomit by a young child in the evening may indicate no more than that the mother had not recognized that he was ill and had given him the usual supper At the onset of high fever when any older child may vomit an infant may have a convulsion and an adult a rigor More persistent vomiting in the presence of fever and in the absence of much abdominal pain must raise the possibility of meningitis Frequent vomiting starting after the child has been to sleep in the absence of fever or other abnormal signs between bouts of vomiting permits the diagnosis of winter vomiting disease to be considered even at other times of the year but all other possibilities such as food

or other poisoning or abdominal emergencies must first be excluded particularly in a solitary case Vomiting is a constant early feature in infective hepatitis in children but this is not necessarily so in adults

The term '**diarrhoea**' often means different things to the doctor and to the patient or his relatives It is therefore safer if in doubt about the value of a negative answer to enquire separately about the looseness and the frequency of the motions Even the occurrence of diarrhoea may not be correctly interpreted The writer has seen an infant become severely dehydrated before the mother or the doctor realized that its nappies were wet by the pale watery stools of a fatal gastro enteritis rather than by urine Diarrhoea alone in a school child at any season but particularly during winter and spring is suspicious of Sonne dysentery Simultaneous vomiting and diarrhoea suggests a severe gastro enteritis due either to infection (though often no bacterial pathogens may be isolated) or to staphylococcal poisoning

Sore throat is seldom complained of by a child under school age even when the tonsils are acutely inflamed though he may admit to it if asked Older children and adults can sometimes distinguish between a throat that is sore all the time but relieved by swallowing and a throat that is painful on swallowing The former sometimes is the case in influenza or adenovirus infection the latter in tonsillitis glandular fever or quinsy A painful pharyngitis may be one of the earliest symptoms of measles In early diphtheria there may be little complaint of sore throat

The commonest reason for a child to wake in the night **crying** is a nightmare especially in children with respiratory obstruction such as those awaiting removal of tonsils and adenoids or a baby with its first head cold In a feverish child pain from acute otitis media is perhaps the next common cause of crying in the night If these conditions can be excluded and especially in the presence of other suggestive signs a night of whimpering and restless crying should at once arouse suspicions of meningitis

Loss of appetite is perhaps one of the earliest symptoms of fever particularly of infections in the nose and throat It is not however proportional to the height of fever many children with high fever never lost their appetite during the Asian influenza

epidemic in 1957 Anorexia with nausea is a presenting symptom in nearly every case of infective hepatitis

Sneezing and coughing occur at the onset of many acute febrile respiratory infections In influenza and in many unnamed short catarrhs sneezing diminishes on the second or third day as the fever subsides in measles coryza increases during the few days which elapse between the initial fever and the appearance of a rash In influenza unlike the short catarrhs coughing is usually painful from the onset Increasing cough in an otherwise well child without sinus tenderness and without signs in the chest should be considered as whooping cough until this can be ex cluded the almost universal use of prophylactic vaccines has made this diagnosis no easier than before

Aching in the body and limbs is an adolescent and adult symptom seldom complained of by a young child even with influenza It is a symptom of poliomyelitis which is becoming familiar to the public Joint and limb pain is often a prominent early symptom of undulant fever differing however from the acute pain and stiffness (not ache) described by an adolescent with acute rheumatic fever

Abdominal pain with or without vomiting occurs in Bornholm disease infective hepatitis and the pancreatitis of mumps as well as in simple gastro enteritis or appendicitis In taking the history it should be noted whether pain preceded vomiting (as is usual in acute appendicitis and colic) or coincided with it (as in winter vomiting disease) The abdominal discomfort of early typhoid fever rarely amounts to severe pain and is often only one of many confusing symptoms

THE SEARCH FOR PHYSICAL SIGNS

Every examination of a patient with pyrexia of unknown origin should follow a methodical routine designed to reveal any abnormalities present According to the patient s age and intelligence greater or less help can be gained from a careful enquiry into the history and symptoms In young children how ever physical signs assume particular importance in guiding one to the correct diagnosis before the clinical picture has fully

unfolded Special attention therefore will be given to the examination of a feverish child while noting differences in physical signs which arise as a consequence of age

Before the young patient is disturbed certain impressions may be formed on entering the sick room One may notice the smell of ketones if the child has been vomiting much others are prone to ketosis as a complication of fever Ketosis is however uncommon early in influenza or the summer pyrexias with headache The dry irritating cough of early measles differs from that of acute chest infections In good daylight there is commonly a pink suffusion of the bulbar conjunctivae in influenza measles and many other virus infections whereas the absence of such reddening once put me on the alert in an early case of scarlet fever during an influenza epidemic The restlessness and crying of a child with meningitis will also be noticeable even before one uncovers or touches the patient

Physical examination especially of a baby asleep in its cot, should begin with counting the pulse at the superficial temporal artery before the child awakens no later record will be more valuable and a good reading of the pulse rate will become impossible after the child starts to cry The doctor therefore needs to have a watch with an easily seen second hand preferably one which turns the full radius of the watch face

Each doctor will evolve his own method of completing the full examination of a feverish child and only experience gained by long practice will teach him when to cut short, or when to extend the search for abnormal signs If possible the writer looks next at the ear drums to exclude acute otitis media since even a timid child will usually allow this to be done as it lies in bed The tonsillar glands are then palpated for enlargement and tenderness If they are not enlarged the fever is probably not due to bacterial tonsillitis whatever the appearance of the fauces If the glands are enlarged whether tender or not the nose and sinuses ears and throat must be searched for evidence of present or past infection Tenderness of the post cervical post auricular or occipital glands is of diagnostic value in rubella and suggestive in other respiratory infections of virus origin such as measles and influenza but palpable enlargement of these glands without

tenderness is not of diagnostic help because such enlargement is present in an increasing proportion of children between infancy and school age even when they are quite well

If the child is co operative I examine the palate and fauces before uncovering the body otherwise this is postponed The palate should be seen by daylight whenever possible since the colours change in electric light In scarlet fever the soft *palate usually shows a macular rash for some hours before* any rash appears on the skin In measles there may be little or nothing abnormal to find on the palate or along the gums at the first examination but the daily changes at these sites and especially the appearance of Koplik s spots make an early diagnosis possible before the rash appears During influenza epidemics many children who complain of sore throat show a granular erythema of the soft palate with an occasional small macule especially on the uvula A pale soft palate and uvula with no abnormal signs in the ears and chest is the usual finding in many undiagnosable fevers especially in summer but in infections by strains of adenovirus and Coxsackie virus as well as in poliomyelitis there may be reddening of the posterior margin of the soft palate Follicular exudate may occasionally appear on the surface of the tonsils without bacterial pathogens being detectable A streak of mucopus on the posterior wall of the pharynx early in the illness suggests that the cause is a bacterial rather than a viral infection

In every case of pyrexia with respiratory symptoms where the diagnosis is still in doubt the absence of abnormal signs in the lungs should be looked upon as a positive rather than a negative contribution suggesting that the illness is due to a general rather than a local infection Auscultation of the chest gives an opportunity to look for a rash on the abdomen and back especially on the pale skin of the axillae and upper arms the flanks and buttocks *The axillae and groins should also be palpated for* tender enlargement of the lymph glands While listening to the back of the chest with the child sitting up the writer tests the neck and back for muscular spasm which is often present in a mild but worrying degree whenever the temperature is over 103 F with lower fever this sign increases in importance as

evidence of meningeal infection Since a rigor high fever and vomiting or nausea either in children or adults is often due to a urinary infection examination of the abdomen should include bimanual palpation of the loins for renal tenderness Tenderness over the appendix or the mesenteric lymph glands the presence of an enlarged liver or palpable spleen or of faeces in the descending colon should be looked for constipation not infrequently causes low fever and abdominal pain

In three different children the writer first recognized the presence of a congenital heart lesion when their pulse rates were quickened and their cardiac sounds accentuated by fever The mothers of all such children should be advised to inform any doctor who has to see the child in the future that such abnormalities were known to have been present in the past lest his finding of them should suggest the early diagnosis of some other illness

Fever may be due to a local infection of the limbs though seldom in a child now that osteomyelitis has become so rare In older patients one still comes across an occasional erysipelas or cellulitis of the leg or foot Gross abnormalities of muscle power in the limbs will be quickly noticed while making a child bend and straighten his knee under the guise of examining for articular abnormalities If there is any likelihood of poliomyelitis a more careful test of the strength and movements of both arms and legs must be carried out There need be no fear of making the parents anxious by this careful examination for the fear of polio is often uppermost in their minds Where this is so and one can be reasonably certain that the child is not suffering from that disease one may decide to say so explicitly

In a dispensing practice such as the writer s it is easy to arrange that the parents bring a specimen of the child s urine for examination when they collect his medicine at the surgery later in the day If the new call comes in by telephone it is better still to ask for a specimen of urine to be kept for the doctor to see at the time of his first visit

The relatives should be encouraged to take the patient s temperature before each meal time three or four times a day and to keep a written record This may be invaluable if the

tenderness is not of diagnostic help because such enlargement is present in an increasing proportion of children between infancy and school age even when they are quite well

If the child is co operative I examine the palate and fauces before uncovering the body otherwise this is postponed The palate should be seen by daylight whenever possible since the colours change in electric light In scarlet fever the soft palate usually shows a macular rash for some hours before any rash appears on the skin In measles there may be little or nothing abnormal to find on the palate or along the gums at the first examination but the daily changes at these sites and especially the appearance of Koplik s spots make an early diagnosis possible before the rash appears During influenza epidemics many children who complain of sore throat show a granular erythema of the soft palate with an occasional small macule especially on the uvula A pale soft palate and uvula with no abnormal signs in the ears and chest is the usual finding in many undiagnosable fevers especially in summer but in infections by strains of adenovirus and Coxsackie virus as well as in poliomyelitis there may be reddening of the posterior margin of the soft palate Follicular exudate may occasionally appear on the surface of the tonsils without bacterial pathogens being detectable A streak of mucopus on the posterior wall of the pharynx early in the illness suggests that the cause is a bacterial rather than a viral infection

In every case of pyrexia with respiratory symptoms where the diagnosis is still in doubt the absence of abnormal signs in the lungs should be looked upon as a positive rather than a negative contribution suggesting that the illness is due to a general rather than a local infection Auscultation of the chest gives an opportunity to look for a rash on the abdomen and back especially on the pale skin of the axillae and upper arms the flanks and buttocks The axillae and groins should also be palpated for tender enlargement of the lymph glands While listening to the back of the chest with the child sitting up the writer tests the neck and back for muscular spasm which is often present in a mild but worrying degree whenever the temperature is over 103 F with lower fever this sign increases in importance as

scarlet fever but in such patients the throat should be swabbed if any real doubt exists Transient rashes mainly on the trunk are not infrequent in glandular fever and some types of virus meningitis and are occasionally present in influenza roseola infantum probably accounts for at least some so called teething rashes

THE INFLUENCE OF AGE

As we grow older we acquire immunity to many infections The chills of infancy and childhood no longer break down our resistance or if they do our reactions to the infection are less violent Do not expect a temperature of 105 F in a grandfather with influenza nor be surprised at finding it in a school child with measles In infants and the aged or infirm however toxaemia may overwhelm the body before its defences can be mobilized

Because the chance of encountering different infectious diseases varies according to where and how one lives and works so the commonest age for contracting measles is higher in the country than in cities Some diseases are demonstrably more infectious than others to those who are equally exposed in schools shops cinemas or buses For these two reasons one finds that in cities measles attack the pre school child but is infrequent after the age of ten scarlet fever is commonest in the middle school years while rubella mainly affects school children and young adults rather than infants But in a country practice the writer has attended a woman of seventy with mumps and a man of over seventy with his first attack of measles while whooping cough has been described elsewhere in old people over eighty A doctor must always consider the bearing of age upon the likely diag nosis but he must never let it over rule his clinical findings

SEASONAL PREVALENCE

Let us now look more closely at the influence of the seasons upon the prevalence of infectious diseases in Britain The fortieth week at the start of October seems in most years to be a turning point The peak of a poliomyelitis epidemic will probably have passed during September and with this will come the end of

illness turns out to be a long one like glandular fever or undulant fever it can be of help in the early diagnosis of measles when the catarrh increases though the temperature drops before the rise which accompanies the appearance of the rash In some diseases the height of the initial fever is a characteristic which may assist in making an early diagnosis while in influenza a second rise of temperature on the fourth day to a level usually lower than that of the first peak will help to confirm a tentative diagnosis Absence of pyrexia is a reliable feature of early *uncomplicated whooping cough Though high initial fever is* common in scarlet fever measles bacterial meningitis Bornholm disease influenza and smallpox a lower temperature does not necessarily exclude any of these conditions In contrast it is unusual for a temperature above 103 F to be found at the first visit for chickenpox infective hepatitis typhoid in the first few days of illness mumps rubella undulant fever or diphtheria When considering the presence or absence of fever at the time of the first visit, one must bear in mind the prevalent habit of dosing everyone who has a headache or seems out of sorts with aspirin or some similar tablet During the 1957 influenza epidemic it was not an uncommon finding for the pulse rate of a child to be over 100 but the temperature to be below 98 4 F Enquiry usually showed that an antipyretic had recently been given which had lowered the temperature more than it had slowed the pulse rate After a few hours without drugs the temperature rose to its proper level often without much rise of pulse rate

The finding of a rash by the patient his attendants or the doctor always heightens excitement at least for a while Because it often appears without much constitutional disturbance the rash of rubella is encountered in a doctor s surgery more often than *those of measles chickenpox or scarlet fever If a morbilli* form rash fades without a firm diagnosis being made the absence of staining in the skin tends to exclude measles in a child under two both measles and rubella are rare unless there is an epidemic in the district while the interval between a primary and secondary case can sometimes be used to make a retrospective rather than an early diagnosis The absence of a rash on the palate in the presence of a punctate rash on the trunk tends to exclude

disease psittacosis cat scratch fever toxoplasmosis or infections by *Pasteurella septica* These diagnoses are only made with the help of a pathologist.

EARLY COLLECTION OF MATERIAL FOR SUBSEQUENT LABORATORY INVESTIGATION

The causative agents of an increasing number of infectious diseases can now be isolated on tissue-culture Early collection of suitable material for laboratory studies should not be forgotten in appropriate cases of pyrexia of uncertain origin This applies to influenza poliomyelitis aseptic meningitis Bornholm disease and other infections by strains of Coxsackie Echo or adenovirus groups as well as to many diseases caused by bacteria. Such material should be collected at the earliest stage in which the patient is seen This applies to all virus diseases though in polio myelitis and Bornholm disease the virus may still be detectable in the stools after more than two weeks

Throat swabs sputum gargle media or faeces from which an attempt is to be made to isolate virus should be frozen solid as soon as possible this does not spoil them for bacteriological examination if required When carbon dioxide snow cannot be obtained in sufficient quantity salted ice in a wide mouthed thermos flask can provide a useful alternative Vacuum venules are more convenient than separate syringes and containers for the collection of paired samples of serum during the acute and convalescent stages of illness if white cell counts are not required Where an attempt is to be made to identify the virus serologically it is usually not too late to collect the acute specimen on the third or fourth day of fever the convalescent specimen may generally be collected between the fifteenth and thirtieth days

In those countries where malaria is a possibility collection of a thick and thin blood smear should be made at the first visit. If necessary anti malarial treatment can then be started without losing the evidence on which a positive diagnosis can later be made

In most bacterial infections diagnostic material can safely be posted to the laboratory without loss of viability In doubtful

a season of Bornholm disease and other Coxsackie virus infections and a diminishing risk of virus meningitis October sees a fresh start of acute chest infections and acute otitis media and the incidence of measles whooping cough and scarlet fever begins to rise By November occasional cases of rheumatic fever may be seen and the winter vomiting disease becomes prevalent By December measles may be widely epidemic spreading to country districts around the large towns and the first wave of Sonne dysentery in schools reaches its peak

After the Christmas festivities with the fatigue and infinite possibilities for cross infection January brings a rising incidence of influenza and perhaps cases of mumps and bacterial meningitis The winter vomiting disease reappears after a short lull while in February or March the main peak of many other winter epidemics is reached influenza whooping cough and measles in turn make way for Sonne dysentery and meningococcal infection With the coming of April Bornholm disease which has occurred sporadically throughout the winter months begins once more to be epidemic May brings its crop of pink eye cases some of which will be due to adenovirus infection In May or June the incidence of rubella which has been rising since New Year reaches its peak while that of measles dies away except in isolated communities among which summer epidemics are not uncommon From July onwards one begins to study every case of fever anxiously lest it should be the first in an outbreak of poliomyelitis Some of these cases of pyrexia of uncertain origin many of them with headache will be due to virus meningitis (possibly one of the Echo or Coxsackie viruses) while others in which sore throat is more prominent may be associated with strains of adenovirus

And so the turn of the wheel is complete At any time of the year however this apparently smooth progression may be disturbed by an epidemic of chickenpox or glandular fever neither of which appears to be as confined as other infections by the seasons or there may be sporadic cases of diphtheria undulant fever infective hepatitis typhoid or paratyphoid malaria or smallpox Even this list does not include other rarities which may never be seen in a lifetime such as Weil s

indigestion or vomiting it was not until later that evening that the friction rub of Bornholm disease could be detected A child of two with acute follicular tonsillitis and tender enlargement of his tonsillar glands should have responded to treatment (at least with a lowering of the temperature) after forty eight hours on parenteral penicillin so the writer applied a tuberculin patch test a positive reading forty eight hours later put him on to more than a dozen cases of tuberculous adenitis spread by a common milk supply

DECIDING FACTORS

A rash may not always appear in measles nor need everyone with mumps show parotitis Formes frustes or atypical cases may be seen in most epidemics Their early differentiation at the bedside with help where applicable from the laboratory depends upon knowing the salient features of a typical early case

Measles

The epidemic season is December to June Rare under six months the common age group is one to ten years The earliest symptoms are sneezing then cough and reddening of the eyes The throat may be sore These symptoms precede the rash by three or four up to seven days Initial fever may be high falling until the rash appears when it rises to 103 105 F Koplik s spots on the buccal mucosa may precede the rash by up to seventy two hours The rash appears first on the face and takes thirty six to forty eight hours to develop fully as far as the legs Atypical cases show coryza and other initial symptoms occurring at the appropriate interval after the primary case but no rash appears and Koplik s spots are rare Prodromal rashes during the catarrhal stage are commonly mistaken for scarlet fever No help can be gained from laboratory tests

Rubella

Epidemics occur in the first half of the year especially in May and June The serial interval between cases in contact is seventeen to eighteen days slightly longer than the twelve to fourteen days in measles It affects school children or young

cases of scarlet fever it is wise to take a nasal in addition to a throat swab the former should be moistened in boiled water before use to diminish the discomfort of this procedure

ASSESSING THE EVIDENCE

While one is taking a history and during examination of the patient the value of each piece of positive or negative evidence is judged against certain background information such as the season of the year and the age of the patient Thus a high temperature and aching limbs in a business man in winter time may start the doctor thinking about influenza whereas the same symptoms in a young athlete after August Bank Holiday are more likely to raise suspicions about poliomyelitis or in a teenage schoolboy on a damp November evening of rheumatic fever Closer enquiry into the history careful consideration of the other physical signs and the pattern of fever as it unfolds however together with one s knowledge of the patient s recent activities or other personal details may lead one in any case to have samples of blood tested for typhoid undulant or glandu lar fever

The ability to make these mental assessments quickly and accurately is one of the richest rewards of well used experience Yet in dealing with infectious fevers in general practice one must be constantly on guard against that familiarity which if it does not breed contempt may blunt one s sense of the inappropriate By this the writer means that *if a symptom or physical sign does not match one s preconceived idea about the diagnosis it is more likely that the diagnosis and not the finding is wrong* Always pay the greatest attention to that part of the clinical picture which will not fit conveniently into one s diagnostic pigeon hole For example a man of sixty five with acute upper abdominal pain tenderness and fever of less than 100 F also had a small palpable sub maxillary gland but it was several days before his parotitis appeared A housemaid who dropped a tray of morning tea on the stairs as she collapsed with sudden severe upper abdominal pain had a temperature of 103 F half an hour later with no other abnormal physical signs and no history of

evidence of sinusitis increasing cough which persists for more than a week and is aggravated by laughing crying and eating is typical of the early stages of pertussis Unlike the cough of a catarrhal child which gets worse on going to bed and getting up whooping cough is increasingly troublesome during the night waking the child with a fright Otitis media and pharyngitis are usually absent there is no fever and no response to penicillin or ordinary cough mixtures At this stage a cough plate which reaches the laboratory within a few hours of exposure may be positive and clinch the diagnosis but a negative finding which is not infrequent does not exclude pertussis A per nasal swab from the posterior wall of the naso pharynx may show a positive result if taken after a bout of coughing even though negative at other times The characteristic lymphocytosis which develops during the paroxysmal stage seldom appears in the first week of the disease With the onset of vomiting after a week or ten days of increasing cough the diagnosis becomes certain even though no whoop occurs Immunisation during infancy alone may not protect fully during the later school years A child partially immunised by vaccine may have a mild attack of less than three weeks duration and yet may spread virulent pertussis to an unprotected infant

Chickenpox

There is no regular season of maximum prevalence The disease may occur in children ten to sixteen days after the appearance of herpes zoster in an adult Mild cases may have less than a dozen spots those severely affected may have over 600 The first lesions usually appear on the chest or flank preceding the second crop of spots by eighteen to twenty four hours Early lesions in each new crop are macular Blistering does not occur in less than about six hours though it can be seen with a lens starting in lesions which appear to be macular to the naked eye and in the prodromal rashes which are not uncommon A doctor is usually called after the second or even third crop of spots has appeared lesions of each crop will then be at different stages of development They are more numerous on the back than the front of the body and on the upper half of the face

adults more often than infants and is infrequent over the age of thirty even among family contacts There may be no prodromal malaise and the rash which first appears on the face or neck spreads to the whole body within twenty four hours fading as quickly with little or no staining again in contrast to measles Fever is commoner in adults than in children Tender enlargement of the lymph glands in the postauricular and occipital groups in the absence of scalp sepsis is an early diagnostic sign which may precede the rash by a day or more Atypically the disease may occur without a rash There are no confirmatory tests

Scarlet Fever

The disease may become locally epidemic at any time of the year but is commonest in spring and autumn Rare in infants the disease affects children in the middle school years and is uncommon after the age of ten differing here from acute rheumatic fever where the main incidence is about puberty The onset of illness is shown by fever sore throat or vomiting either singly or in combination according to the age of the patient The pulse in early stages of scarlet fever is disproportionately fast Earache may be present at onset as an early complication The palate or anterior pillars of the fauces are usually reddened and macular erythema often appears on the soft palate several hours before the rash develops on the trunk The rash avoids the circumoral skin which appears pale in contrast to the flushed cheeks Atypical cases may show little or no tonsillitis but enlargement of the tonsillar lymph glands with or without tenderness is always present A dry swab of the fauces and a wet swab of the nose should be taken in any case of doubt *Group A beta haemolytic streptococci* can be cultured either from the throat or nose or—in atypical cases—from a skin lesion elsewhere on the body such as an infected graze on the leg

Pertussis

Epidemics of this disease are becoming uncommon except in unvaccinated families Spring and autumn are the usual seasons for prevalence In a healthy child without signs in the chest or

Infective Hepatitis

The season of main prevalence is not well established and local epidemics may occur at any time The serial interval between cases is three to five weeks The disease may affect any age group but the youngest are more susceptible Preceding jaundice by three to six days the early symptoms are fever with headache anorexia nausea and abdominal discomfort or pain the patient feels unwell and an adult may lose his taste for tobacco Pale stools and bile in the urine appear one or two days before jaundice Atypical cases occur in which clinical jaundice is absent in these the wheal produced by an intradermal injection of 0 25 mg of histamine is discoloured yellow Blood examination is also helpful in doubtful or prolonged cases of jaundice In an atypical case of febrile jaundice in a young adult a positive Paul Bunnell reaction may indicate that the correct diagnosis should be glandular fever

Glandular Fever

The season of maximum prevalence has not yet been established The incubation period in volunteers is five to twelve days but some serial intervals between naturally occurring cases in family outbreaks are as long as one month Children and young adults are mainly affected in adults the course tends to be more prolonged Since the syndrome of glandular fever is not uniform the diagnosis is often reached only by exclusion The sudden onset of fever with slight sore throat and adenitis which increases for a few days without much tenderness are characteristic of the disease in children or adolescents enlargement of the sub sternomastoid lymph glands is almost pathognomonic There is no response to treatment with penicillin In adults the disease may present with increasing fever and headache malaise sweating and muscular pain but with scarcely any sore throat A week later a transient maculopapular rash may appear on the trunk and limbs for three to five days and not until after that may the glands enlarge Throat swabs taken in anginose cases may show a variety of organisms but few pathogens and diphtheria bacilli are absent even when membrane is present on the tonsil Among other atypical

High fever is rare commonly there is none when the first spots appear Herpes zoster and chickenpox can be present con currently in atypical cases and the writer has seen a child have a second attack of chickenpox within six weeks Confirmatory tests carried out on vesicle fluid are rarely required

Mumps

The epidemic season is winter and spring All ages can be affected but the disease is rare in infants and the aged being commonest in school children A short prodromal fever may occur a few days before any swelling appears While the virus commonly infects the salivary glands these may escape altogether The first evidence may then be either orchitis encephalitis or—rarely—pancreatitis Acute labyrinthine dis turbance may be an early or even a solitary symptom Any patient without salivary gland enlargement who shows one of these manifestations should have complement fixation tests for mumps done on specimens of his blood taken during both the acute and the convalescent stages of illness

Influenza

Epidemics commonly begin early in the New Year but—as in 1957—infection may be introduced at any season and spread rapidly if resistance is low The main incidence is upon youth and middle age rather than upon infants or the aged but these fare badly when affected The onset is usually abrupt with headache a painful cough aching limbs and a high or mounting fever as the early symptoms Sore throat may be a feature among children In fulminating cases hyperpyrexia and death from toxaemia or heart failure may occur within twenty four to forty eight hours of onset Early death may also occur in those with pre existing heart or lung disease Laboratory confirmation can come from complement fixation tests on paired sera taken during the acute and convalescent stages or from isolation of the virus on swabs of the nose or throat or in sputum especially if the material is collected when the fever reaches its peak and is quickly frozen Atypical cases confirmed serologically may show malaise with little or no fever and yet be infectious to others

after the bouts of pain is one of the few physical signs present in most patients rarely a friction rub will be heard near the site of pain arising not in the pleura but in muscle fascia Atypical cases may show marked meningeal symptoms and signs which only lumbar puncture and laboratory tests can distinguish from septic meningitis when they occur sporadically Mild afebrile cases are common during epidemics Virus can be isolated from the majority of patients if the stools are examined twice during the first two weeks of illness Serological tests are possible but are not carried out as a routine

Poliomyelitis

This is dealt with separately in Chapter XIII

PYREXIA OF UNCERTAIN ORIGIN

Sydenham also remarked that as to Fevers we must take notice that the greatest part which are continual have yet no Name allotted to them Osler in his day divided the remnant into three categories acute catarrhal fever ephemeral fever lasting only about twenty four hours and febricula in which the temperature was raised for three or four days but without localizing signs At the present time and with increasing aid from the laboratory it is probably true to say that a name can eventually be given to nearly all fevers lasting over a week though it may take more than that time to establish a diagnosis in some of these patients As regards Osler s ephemeral fevers however we have made but little progress and we should be as ready as Sydenham and Osler to acknowledge that we do not know their causes

An entry in one s notebook P U O—rash and teething is just as scientific as and more honest with ourselves than the entry Teething rash I make a point of recording the prominent symptoms or signs in all cases of pyrexia of unknown origin hoping that one day as I go back over these notes I shall recognize in retrospect that the patient was suffering from some as yet unidentifiable infection After learning to recognize Born holm disease I well remember the thrill of turning through an

features are jaundice and neurological manifestations Abnormal lymphocytes and monocytes appear in the blood when glandular enlargement occurs or soon afterwards the total leucocyte count usually lies between 10 000 and 20 000 per cm The Paul Bunnell reaction does not usually become positive until the end of the pyrexial stage in children therefore it may be positive at the time of the first visit while in adults it may not appear until a fortnight or more from the onset of symptoms In many cases of clinically typical glandular fever the test remains negative throughout the illness

Bacterial Meningitis

This is mainly a disease of infants and young children during winter and spring Sporadic cases are commoner than epidemics the latter when they occur are usually confined to young adults living in over crowded quarters The disease starts as an abrupt fever with few localising signs Headache restlessness and vomiting are early symptoms in children and adults infants may not vomit but have one or more convulsions instead Photo phobia dorsal spasm and rash do not appear early Recovery and specific treatment based on a correct diagnosis require the early performance of lumbar puncture Atypical cases showing only naso pharyngeal symptoms can be detected among contacts by swabbing

Bornholm Disease

Sporadic cases are commoner than multiple and may be seen in any month of the year epidemics usually occur between April and October Children under fifteen are principally affected but the disease also attacks adults of all ages Severe pain localised to the lower chest and upper abdomen coming on rapidly and aggravated by movement or breathing but not shift ing its position is the principal feature The pain after lasting for several hours often ceases abruptly High fever coinciding with the attack of pain and abating as the pain fades is also characteristic but many mild cases have little or no fever One or more relapses of pain and fever may occur at intervals of twenty four to thirty six hours Muscle tenderness during and

CHAPTER IV

MALIGNANT DISEASE OF THE ALIMENTARY TRACT

By Ian Aird

CARCINOMA OF THE STOMACH

SOME arrangement for men and women at and beyond middle age to have a physical examination every six months or so in the hope of early discovery of malignant disease has many superficial attractions In practice there would be some objections to it Were only carcinoma of the stomach to be considered a barium meal examination performed on apparently healthy subjects every six months after the age of fifty would be within the limits of the possible if expense were no object Some un suspected carcinomas would be found but it would require many thousands of barium meal examinations to furnish a single positive diagnosis The cost of discovering one carcinoma of the stomach in this way might well amount to a quarter of a million pounds for each positive radiological diagnosis The demonstration of a very early lesion and the close examination of a doubtful condition require much more care and time from the radiologist than the demonstration of a florid lesion and it is doubtful whether we have sufficient radiologists and radiographers available in the country for a large scale attack on the problem in this way If this kind of examination referred not only to the stomach but also to the colon and to some other parts of the body where malignant disease is common the project would assume vast proportions if it were applied on a national scale Nor would all the lesions detected by this kind of technique prove curable by surgery

There is however one simple kind of examination which might be used as a first filter Most though not all patients who

old notebook and finding the diagnosis for a youth entered one April as acute diaphragmitis—? cause

Without more close clinical study particularly by general practitioners and more research particularly in virus laboratories many of our patients in whom the diagnosis starts as pyrexia of uncertain origin must be content with an ultimate diagnosis of pyrexia of indeterminable origin Osler s diagnosis of febricula was shorter and just as accurate

The first part of the clinical investigation of a case in which cancer of the stomach is suspected undertaken in the consulting room is a complete physical examination The abdomen is carefully palpated for tenderness and for swelling in the first place There is no tenderness in many carcinomas of the stomach but some—especially of the ulcerative varieties—are tender and tenderness in the epigastrium of a patient in or beyond middle age justifies further investigation The presence of a tumour is of course important if one can be felt and its mobility should be tested Even a large palpable tumour should not give rise to despair for surgery finds some of its greatest successes in the removal of large glandular gastric neoplasms If a gastric cancer has grown to a large size without producing demonstrable metastases and without killing the patient it may well be of an inherently low order of malignancy

Of course even though the symptoms have been of the slightest it may be already obvious at first examination that the patient has passed beyond the possibility of surgical cure There may be free fluid in the abdomen or large palpable peritoneal masses or enlargement of the liver or palpable glands above the clavicle or a thickening of the pelvic peritoneum palpable on rectal examination Even then though cure may be impossible the patient should still be offered the opportunity of exploration for gastrectomy will often prolong life and can make survival more tolerable

The stool should always be examined for occult blood in any case where there is a suspicion of cancer of the stomach A negative result does not exclude the disease but a positive result makes further investigation obligatory

Carcinoma of the stomach cannot be excluded without the performance of an opaque meal examination I believe that the omission of this kind of examination is permissible only if there is a palpable tumour in a middle aged patient such a finding means that the patient must in any case come to surgical exploration and for reasons of economy it seems justifiable to omit an expensive and time consuming form of investigation and not to delay an inevitable operation for this to be done

suffer from an early carcinoma of the stomach have occult blood in the stools and a patient who insisted on a periodic check might well have his stool tested for blood If this proved positive—and the technique is a simple one—further investigations could then be undertaken

Positive physical signs should not be forgotten in this connection Sometimes a cancer of the stomach turns up unexpectedly during routine physical examination of the abdomen Sometimes a cancer of the stomach which grows to a large size before it produces symptoms proves highly amenable to surgery and many surgical successes are obtained by the operative removal of large glandular tumours If every patient who presents in the consulting room is subjected to a complete physical examination an occasional gastric tumour will crop up even in the absence of all gastric symptoms

In considering early symptoms it is important to remember that cancer of the stomach does not always first present as a form of dyspepsia Nevertheless the very slightest symptom of indigestion in a patient in or beyond middle age should lead to a full gastric investigation These symptoms may be quite inconsiderable and they include diminution of an appetite previously normal a disinclination for food after the first few mouthfuls a feeling of slight fullness after a meal epigastric discomfort or even unaccustomed belching regurgitation or a bad taste in the mouth Actual epigastric pain sometimes quite severe and suggestive of ulcer is not a common presenting feature of carcinoma of the stomach but it is occasionally the first symptom Acute epigastric pain can be a symptom of cancer

More commonly the first symptoms of carcinoma of the stomach are not noticeably gastric There may be loss of weight noticed by the patient or his wife as a sole symptom A general feeling of weakness sometimes accompanied by a feeling of lassitude and lack of energy and sometimes proving in due course to be associated with a secondary or even a primary anaemia should raise the suspicion of cancer of the stomach in a doctor s mind even if there are no dyspeptic symptoms

Gastroscopy is seldom essential for the diagnosis of malignant disease of the stomach It is particularly useful in differentiating a simple ulcer from a malignant one though it cannot be always fully depended upon even for that and it is useful to distinguish the unbroken mucosa over a simple polypoid tumour from the ulcerated surface of the typical proliferative carcinoma

Gastric washings can be spun down and examined histologically after suitable staining for malignant cells The histological technique is a specialized one and experience is required before a precise and reliable diagnosis can be achieved but in the hands of a practised team the method is of additional advantage in doubtful cases

Sometimes a cancer of the stomach first attracts attention by perforating Those cancers which perforate are in the main ulcerative types of malignant disease The abdomen is opened usually under a diagnosis of perforated peptic ulcer and such an ulcer is found often with little to suggest that it is carcinomatous A small biopsy should always be taken when a perforated gastric ulcer is closed particularly in a patient of or beyond middle age

Attention has recently been drawn to a relationship between gastric cancer and pre existing atrophic gastritis and some American authors are now accustomed to suspect malignant disease in stomachs shown at gastroscopy to be the seat of the atrophic lesion Some such patients do in fact prove to have early carcinoma in the atrophic mucosa but in Britain prophylactic resection is not commonly undertaken in these circumstances

CARCINOMA OF THE DUODENUM

Carcinoma of the duodenum is an extremely rare form of malignant disease When it does occur it affects the second part of the duodenum in the neighbourhood of the ampulla of Vater It causes jaundice early and it behaves less like a carcinoma of the alimentary tract than as a carcinoma of the bile duct or of the ampulla of Vater The rare carcinomas of the lower duodenum behave for all practical purposes as carcinomas of the small intestine

The radiological pattern produced by a cancer of the stomach is very variable and early abnormalities may be very slight A localized disappearance of gastric folds is suspicious and localized rigidity of the gastric wall under palpation may be the first radiological sign Later there may be a filling defect This may take the form of a circumferential stenosis or of an intrusion into the lumen from one diameter, or of an irregular island or series of islands in the centre of the barium shadow The lesion may occlude the pylorus causing delayed emptying of the stomach usually without much dilatation or a tubular carcinoma may hold open the pyloric canal so that the stomach empties quickly Rarely there is an hour glass appearance with a cup and spill or cascade deformity or in the case of cardiac growth a residue of barium in the oesophagus and a small narrow stomach The leather bottle stomach which often produces extensive infiltration of the stomach wall without much in the way of symptoms gives a changeless outline and an incompressible tube of barium

Displacement of the stomach even if the symptoms have been recent is a discouraging radiological sign for the stomach is not readily displaced by malignant tumours that arise within its own wall Displacement suggests the presence of large glandular masses outside the stomach or an enlarged liver

The radiological discovery of an ulcer in the stomach of a patient in or beyond middle age requires very careful observation and thought Cancer should be suspected particularly if an ulcer crater is seen in an unusual situation In a patient of any age any ulcer of the antrum or of the greater curvature or of the fundus should be regarded most seriously and should be surgically explored Size alone is no criterion of malignancy for though some malignant ulcers are very large the largest of all ulcers particularly those in elderly people are often simple More important than size is a radiological appearance indicating proliferation at the ulcer's edge Only in the case of a smooth and typical lesser curve ulcer is it permissible to watch a patient of cancer age for a period of a couple of months to see the effect of strenuous medical treatment.

shortening in length as it does over a Miller Abbott tube The distance from lesion to bleeding point is thus known and the level of the bleeding point below the duodeno jejunal junction can be estimated This information is helpful at the laparotomy which should follow the detection of bleeding from a small intestine lesion

Not all malignant lesions of the small intestine are carcinomatous Some are due to lymphosarcoma reticulosarcoma Hodgkin's disease or one of the other reticuloses These do not commonly give rise to bleeding but rather to the development of intestinal obstruction the nature of which is discovered at laparotomy

CARCINOMA OF THE COLON

The presenting symptoms of carcinoma of the colon differ a little depending on whether the right or the left colon is the seat of the disease

In any part of the colon an ulcerating lesion can by the frequently repeated loss of unnoticed quantities of blood in the stool give rise to a secondary anaemia with the same feeling of lassitude weakness and disinclination for work or play similar to that so commonly seen in early carcinoma of the stomach As in the case of carcinoma of the stomach the detection of anaemia leads to the finding of occult blood in the stool and if a barium meal proves negative barium enema is undertaken and may reveal a filling defect in some part of the colon This train of events is more common in the case of a tumour of the right or of the transverse colon than of the left colon

Characteristically a tumour of the caecum or ascending colon gives rise to pain and tenderness on the right side of the abdomen On physical examination it is often appreciated that the tenderness is along the line of a palpable bowel and a tumour may even be felt Occult blood may then be found in the stools though not necessarily so the writer has had a patient with a *large malignant ulcer of the ascending colon provide a stool* apparently free from occult blood on thirteen successive days

In any part of the colon of course a malignant tumour may take the form of a string stricture and give rise to acute or

CARCINOMA OF THE SMALL INTESTINE

Carcinoma of the small intestine is a rare disease and it manifests itself in one of three ways It can present as an acute subacute or a chronic *obstruction* it can produce *haemorrhage* or it can remain latent until distant *metastases* in lymph glands peritoneum or liver are already present For our purposes only the first of these presentations are important

It is not very common for a tumour of the small intestine to occasion acute intestinal obstruction for the contents of the intestine are so fluid that they can be propelled past a carcinoma even after the lumen has been very tightly narrowed If the patient does however present with an acute intestinal obstruction no precise pre operative diagnosis is possible operation is per formed and the obstructing tumour is found It is rare for a carcinoma of the small intestine to produce even a subacute or a chronic obstruction but if it does so fluid levels and gas distended loops will be visible on straight X ray If a carcinoma of the small intestine presents early it usually does so by bleed ing Either there is overt melaena or the development of a progressive anaemia In the latter event the presence of intestinal haemorrhage can be proved by the detection of occult blood in the stool When this happens barium meal examination is commonly undertaken and a normal oesophagus stomach and duodenum are demonstrated This is usually followed by a barium enema and this proves to be negative too The small intestine then comes under suspicion and a barium follow through is done The demonstration of a non obstructive lesion in the small intestine by radiological methods is exceedingly difficult and an early carcinoma of the small intestine cannot usually be demonstrated The level of bleeding should then be decided by other means A length of weighted tape is swallowed and left to pass down into the small intestine It is allowed to remain in place for six hours and is then withdrawn and tested by guiac for blood in its whole length The upper limit of bleeding is thus revealed Provided no obstruction is present this gives a fairly accurate indication of the level of the actual bleeding for blood in the small intestine passes distally provided its flow is un obstructed and the intestine does not telescope over a tape

polyposis and the performance of colectomy when polypi can be demonstrated reduces the likelihood of members of that family dying as such patients usually do if they remain untreated from cancer of the colon Similarly the present fashion for colectomy as a treatment for ulcerative colitis saves some of these patients from cancer of the colon which is a risk—though a lesser risk—in this instance also

CANCER OF THE RECTUM

The diagnosis of cancer of the lower half of the rectum is made by digital exploration the diagnosis of cancer of the upper rectum by proctoscopy or sigmoidoscopy Rectal examination proctoscopy and sigmoidoscopy are performed in every case of bleeding from the rectum which is the first sign of carcinoma These examinations are carried out in cases of rectal haemorrhage even if piles are present for piles are sometimes secondary to a cancer of the rectum and they sometimes present coincidentally with that disease Cancer of the rectum can produce the same kind of diarrhoea as does cancer of the colon and when a middle aged or elderly patient has a change in bowel habit the barium enema and sigmoidoscopy which are then obligatory must of course invariably be preceded by rectal examination Nothing so humiliates a clinician as for the radiologist to return to the ward a patient referred for barium enema on the grounds that a carcinoma just inside the anus prevents the barium enema catheter from entering the rectum Carcinoma of the rectum may first present as an acute intestinal obstruction and in every case of acute intestinal obstruction rectal examination should be performed

A complaint of tenesmus and a feeling of incomplete evacuation are fairly common signs of bulky rectal tumours of polypoidal tumours and of tumours just inside the anus

Papillomas of the rectum have a much more malignant character than for example papillomas in the urinary tract It is legitimate in the first place to excise such a tumour but any recurrence should be treated whatever the histology as a carcinoma of the rectum Polypoid tumours of the rectum are less

subacute intestinal obstruction coming to operation with that diagnosis

In the descending and pelvic colon symptoms may be initially slight and they are sometimes not directly referable to large bowel If they direct attention to the colon they should however slight, be seriously treated in a patient of the cancer age Any change in the bowel habit of a middle aged or elderly person manifest in looseness or constipation deserves the closest attention A barium enema should be undertaken and a filling defect or a stricture may be obvious in the X ray plate of the enema, or in the plates taken for contrast after the evacuation In such a case even if the barium enema plates seem to show no abnormality sigmoidoscopy ought to be performed as well It sometimes shows a tumour that is not demonstrable radiologically

The same kind of suspicion should be aroused if any disturbance of digestion in a middle aged or elderly person fails to be explained by a barium meal for the initial symptoms of a carcinoma of the pelvic colon may be referred to the stomach

Sometimes a patient s attention is first attracted to a cancer of the colon by his or her actually feeling an abdominal tumour Such a palpable tumour like some bulky cancers of the stomach is not necessarily hopeless for surgery provided it remains mobile In the colon as in the stomach large polypoid glandular tumours often afford high hope of surgical cure

Sometimes quite a small tumour of the colon too small in itself to occlude the lumen gets caught by peristalsis and passed onward or backward as a small intussusception The patient suffers from attacks of quite severe abdominal colic and if he is kept under observation for a day or two it may be found that a tumour is sometimes palpable and sometimes not This kind of situation is apt to arise both in the transverse colon and in the pelvic colon and a tumour that is sometimes present and sometimes absent should suggest to the clinician an early carcinoma with secondary recurrent intussusception

In the colon as nowhere else in the intestinal tract, there is a possibility of the surgical prevention of carcinoma The careful management of families whose members suffer from inherited

CHAPTER V

HYPERTENSIVE VASCULAR DISEASE

BY JOHN MCMICHAEL

HIGH blood pressure is a physical sign rather than a disease Like other physical signs it will not be found until it is specifically sought Once detected its interpretation requires considerable finesse of judgement and knowledge of the many causative factors that may be involved

DETECTION

The sphygmomanometer cuff which should be as wide as possible should be firmly applied to the upper arm After rapid inflation to above systolic pressure it is allowed to leak until the thud of systolic pressure becomes audible through the stetho scope placed over the brachial artery slow release of pressure is necessary at this point since due to its inertia the mercury column may fail to follow accurately too rapid deflation of the cuff Similarly slow deflation is necessary around the diastolic pressure reading which is taken when the thudding pulse sounds become muffled

Having obtained a pressure reading we must have some idea of where normal begins and ends in various age groups The figures in the table below are averages found in various popula tion studies

Table of Average Blood Pressure

Systolic

Age	*5*	*15*	*25*	*35*	*45*	*55*	*65*	*75*
Male	85	117	124	125	131	146	152	150
Female	85	117	123	128	134	153	172	177

Diastolic pressure rises slowly in each decade from 60 (aged 10) to 85 in males and 95 in females at age 75

dangerous but after their removal too the patient should be followed carefully for years There may be other polyps at a higher level and these like those of the rectum carry some risk of the development of cancer

When a diagnosis of carcinoma of the rectum is made an attempt should be made to carry out a barium enema Some times the rectal carcinoma has arisen in one of several polyps and small polyps higher in the colon are often shown up more clearly by barium enema than by palpation of the colon in the abdomen when the patient comes to operation

the artery is compressed and may yield readings as much as 20 mm mercury too high With a wide firm cuff completely round the arm and firmly applied the error is reduced It should not be forgotten that obese people have in fact a high incidence of hypertension

Some degree of rise in blood pressure as age advances may be accepted as part of the process of ageing The great bulk of hypertensive disease calling for special consideration is that designated *benign* or *essential hypertension* which increases in frequency after the age of thirty and is common in middle life Usually easy to recognize when the pressure falls well outside the accepted upper limit of normal it is unaccompanied by recognizable causative conditions in the kidney or elsewhere In it earliest stages essential hypertension is labile and the pressure usually falls to normal with rest or sleep In a later phase of the disorder the pressure falls under the same conditions of relaxation but does not quite reach normal It has often been suggested that these rises of blood pressure under conditions of stress may be ignored This is not so for what is usually called a normal pressure is a common finding at the time of a medical consultation and those who show a rise under such conditions of stress are in fact abnormal The later evolution of such cases nearly always confirms that they are hypertensive or that they develop arterial trouble in a few years a spit in the wind usually presages rain There are other ways in which this early abnormality may be elicited Immersion of a hand in ice cold water elicits a reflex rise in arterial pressure which is abnormally high in such subjects Desoxycorticosterone acetate (DCA) is also said to induce a pressor response in early hypertension though failing to do so in normals Another indication of their vasomotor instability is a tendency to show abnormally rapid pulse rates at the time of examination (over eighty per minute)

As part and parcel of the undue reactivity of the vasomotor system it is well known that blood pressure will be higher when measured by a strange and white coated doctor than when measured by the same doctor when he has become familiar and in turn it is often lower still when measured by trusted nurses This finding has led to much work on basal casual and

It will be noted that the arterial pressure rises as age advances A great deal of this rise is probably due to ageing processes in the elastic walls of large arteries and thickening of the walls of smaller vessels As the arteries become stiff each stroke volume of blood ejected by the left ventricle causes a more abrupt and larger rise of aortic pressure than would be caused by the same systolic ejection into a more yielding elastic artery This affects particularly the systolic pressure and thus systolic hypertension is often seen in older people The diastolic pressure on the other hand is determined by the run off' from the large arteries during diastole when the arterioles which control this flow are tightly constricted the diastolic pressure will be high Thus a systolic hypertension (*e g* 210/85) usually indicates hardening of the large arteries with advancing years a high diastolic pressure (*e g* 210/125) means a considerably increased resistance to flow through the arterioles The pulse pressure (systolic minus diastolic) depends on two factors the stroke output of the heart, and the rigidity of the arterial wall A low pulse pressure (135/110) usually signifies a falling stroke output at each heart beat while the increased pulse pressure in diastolic hypertension (*e g* 200/110) results from the tighter diastolic distension of the walls of the large arteries this makes the aortic walls more rigid and the pulse pressure rises considerably when the heart thrusts its usual quota of blood into the great vessels

From these principles it may also be deduced that vasodilatation will lower the diastolic pressure and that increased stroke output (such as we encounter in anaemia and early thyrotoxicosis) will tend to raise the pulse pressure In aortic incompetence a large volume of blood is thrust into the aorta causing a large rapid rise of the arterial pressure followed at once by a rapid fall when blood leaks back into the ventricle as well as out into the peripheral arterioles These excessive and rapid volume changes lead to the well known phenomena of the Corrigan pulse

Obesity is a source of error a fat arm may lead to mistakes in the sphygmomanometric reading A good deal depends on the firmness with which the cuff is applied and certain cuffs in common use may fail to surround fat arms A slack cuff expends a good deal of pressure in displacing the subcutaneous fat before

DIFFERENTIAL DIAGNOSIS OF HYPERTENSION

We now recognize a considerable number of types of hyper tension Just as the old pernicious anaemia broke down into a number of varieties of macrocytic anaemia following the precise definition of the haemopoietic factors so we may expect a more precise definition of the hypertensions when pharmaco logical methods of control are more precisely evaluated

Meantime the following associations should be kept in mind

	Before age 30	Nephritis	*Any age*
		Pyelonephritis	Polyarteritis
Cushing s syndrome	Age 30 to 40	Coarctation of aorta	Phaeochromo cytoma
	Age 40 and over	Essential hypertension	Narrowing of the renal arteries
Diabetes mellitus	After 60	Athero clerosis	

RENAL FACTORS

Though there is transient hypertension in acute nephritis eighty five per cent of patients recover completely from this disorder and a history of such an attack is usually of little significance in relation to hypertension in later life On the other hand insidious nephritis (Type II Ellis) often leads to the development of complicating hypertension Nephritis is mani fested by *persistent* albuminuria and usually granular and epi thelial casts are demonstrable in significant numbers in the urinary deposit An occasional hyaline or even a granular cast in the spun deposit is however of little diagnostic significance *Nephritic hypertension is seldom extreme except in the late* malignant phase It is often fluctuating and transient ranging for example from 130/80 to 170/110

Chronic pyelonephritis is probably the responsible factor in about one in six victims of severe hypertension Only about half of these are diagnosed during life A frank history of febrile attacks with renal pain and cystitis is obtained in only about half

supplemental blood pressures the latter being the difference between the first two readings There are difficulties in the interpretation of such measurements since it is difficult to draw the line in the degree of psychological sedation necessary for making the basal reading and the degree of alert awareness on the part of the patient for the elicitation of the casual pressure measurement It has long been known that the physician may vary the blood pressure reading by the expression on his face as he makes the estimation the quick indrawn breath suggesting alarm on the part of the observer has an especially baneful influence

Pregnancy presents special problems The development of even a moderate elevation of pressure is taken as a possible warning of incipient toxaemia Though the vast majority escape without the development of eclampsia this type of hypertension often foreshadows the development of hypertension in a more severe form at a later age even if the pressure appears to return to normal between pregnancies

It is important to keep in mind certain **transient acute hypertensive reactions** which may create clinical confusion Mild hypertension during an acute nephritis may be accompanied by venous pressure elevation and other symptoms of heart failure The picture can be confusing when the albuminuria is minimal as sometimes occurs

During attacks of left ventricular failure from whatever cause (aortic valve disease hypertension ischaemic heart disease) the arterial pressure is often found at a level higher than usual for that particular patient This may confuse the aetiological diagnosis—for example a patient with aortic stenosis whose pressure was 120/85 had a pressure of 180/110 during an attack of dyspnoea The pressure only subsided to the previous level on recovery

Patients recovering from poliomyelitis with bulbar involvement are also said to develop hypertension No doubt this is neurogenic but further studies are awaited

removal of a kidney destroyed by disease and rendered completely functionless is seldom of value in the treatment of hypertension

Polycystic kidneys hypoplastic kidneys renal stones hydronephrosis and polyarteritis affecting the renal vessels may all have to be considered in the renal causation of hypertension

COARCTATION OF THE AORTA

Narrowing of the aortic arch beyond the origin of the left subclavian artery leads to hypertension in the upper part of the body with poor pulse volume and reduced pressure in the femoral arteries The collateral arterial circulation develops and enlarged arteries may be found traversing the lower part of the posterior triangle of the neck medial to the scapulae and in the region of the superficial epigastric arteries of the abdomen Radiological signs include rib notching under the upper ribs where the intercostal arteries are enlarged and tortuous The condition is amenable to surgical correction in early life but the operation becomes more risky after the age of thirty by which time degenerative changes have developed in the aorta The operation reduces the blood pressure but it seldom returns completely to normal

ENDOCRINE FACTORS

Harvey Cushing thought that the disorder which bears his name was caused by a basophil adenoma of the pituitary though it was later recognized that **Cushing s syndrome** was essentially due to oversecretion of hydrocortisone by overactive adrenal glands The administration of hydrocortisone closely reproduces the features of the clinical syndrome

All patients show some degree of hypertension but the other features give the clue Hydrocortisone promotes conversion of protein to carbohydrate and fat Thus there is protein depletion with muscle wasting and weakness the skin is thin and gives way in purplish striae Obesity develops in the face and trunk while *the limbs remain thin The plethoric appearance is based* on a thin skin and the same factor is responsible for the purple colour of the striae Osteoporosis hirsutes gonadal suppression

of the cases The others have a more silent and insidious form of the disorder within the substance of the kidney often lacking the classical recognizable clinical manifestations of the disease In these the presence of variable intermittent and often slight albuminuria with the occasional presence of pus casts may be the only pointers to a correct diagnosis It should be remembered that pus cells are often abundant also in active acute or subacute Bright s disease It is often difficult to be certain of the meaning of bacilluria since contamination is a great bugbear and cystitis without renal involvement common in women More information may be obtained from radiological studies showing shrinkage of one kidney in a plain X ray of the abdomen or patchy reduc tion of renal cortical thickness and distortion of the renal calyces in a pyelogram Needle biopsy of the kidney is helpful if positive but it can be misleading as the lesions are patchy

The role of renal ischaemia in the production of hypertension has attracted much attention in recent years reduction of calibre of one or both renal arteries experimentally or by disease is known to provoke the production of pressor substances by the kidney (Goldblatt kidney) Such lesions occur more frequently in man than has been recognized clinically Abnormal origin and course of the renal arteries may lead to anatomical kinking and partial occlusion in early life while local atheromatous patches with mural thrombosis more often occur in middle or later years Where such lesions are suspected they can only be demonstrated by aortography following the injection of a radio opaque dye (Diodrast) above the renal arteries A slightly less risky method is the demonstration by retrograde ureteric catheterisation of impaired excretion of sodium by the affected kidney in the presence of unilateral constriction of a renal artery

These are no longer impractical matters for removal of a unilateral shrunken kidney may lead to cure of severe hyper tension Even reconstruction of narrowed arteries is a feasible proposition to the modern vascular surgeon Two matters are worthy of mention The likelihood of an underlying renal cause is so high in younger cases that it is the duty of the practitioner to ensure that this possibility is excluded in all patients who are found to be hypertensive under the age of forty Secondly the

ESSENTIAL HYPERTENSION

Essential hypertension is diagnosed when the other factors enumerated above have been excluded It is often said that the *diagnosis of essential hypertension is favoured* by the finding of a family history Studying the inheritance of the disorder Pickering and his associates have found that the siblings and children of hypertensive subjects have on the average higher pressures than normal controls The difference however is slight, and the condition is to be regarded less as an inherited disorder than as a graded characteristic comparable to the inheritance of tallness Galton showed that the heights of the sons of tall fathers fell on the average midway between the heights of the fathers and the average height of the population hypertension in families is somewhat comparable Family history is always difficult to assess It is true that premature death from hypertensive vascular disease in several members of a family may be alarming But those who have long lived parents or grandparents also have a strong family history of arterial disease since it is usually arterial degeneration which finally causes the death of aged people Furthermore nephritic hypertensives have been found to have just as strong a family history as essential hypertensives

Headache is often mentioned as an early symptom of benign hypertension This symptom however is rare in the early stages Sometimes hypertensive headache can be regarded as a manifestation of an anxiety state induced by injudicious reference by the doctor or others to high blood pressure This type of headache will usually be accompanied by other functional manifestations Sometimes migraine may become exaggerated with the development of hypertension When these have been excluded there is a genuine hypertensive headache the features of which are worthy of recognition This headache usually comes on in the morning often awakening the patient from sleep It is severe and frontal or occipital in situation Sometimes it is felt down the back of the neck when it may be sufficiently severe to suggest meningeal irritation Lying in bed helps little but the headache tends to pass off a few hours after rising This type of headache is usually relieved by hypotensive agents and it is

(amenorrhoea impotence) and occasionally glycosuria (25%) are also found

17 hydroxysteroids may be estimated in the urine but the demonstration of a normal value does not exclude the diagnosis Presacral air insufflation may demonstrate a tumour but even in the absence of such findings adrenal overfunction cannot be ruled out The diagnosis of Cushing s disease is essentially made on clinical grounds When ninety per cent of the adrenal tissue is removed under cortisone cover or even when a hundred per cent is excised with subsequent cortisone maintenance the symp toms may be completely relieved

It should be remembered that some plethoric and obese hyper tensive subjects may resemble Cushing s syndrome and care should be taken to ensure that at least a number of the other clinical features are there to support the diagnosis

In the presence of **phaeochromocytoma**—a tumour of the suprarenal medulla—excessive amounts of adrenaline and nor adrenaline are secreted into the blood Paroxysmal attacks of hypertension are characteristic Pallor sweating and palpitation (often slow and heaving) accompany the attacks The thyroid gland swells a little during the paroxysm which may come on during straining at stool Glycosuria is sometimes observed

It should be remembered that fluctuations in blood pressure are common in essential hypertension and unless the other accompaniments suggest phaeochromocytoma the condition will be sought more frequently than it is found. Occasionally how ever phaeochromocytoma may lead to sustained hypertension when the clinical diagnosis becomes very difficult

The tests to be employed are those for estimation of catechol amines in urine This involves biological estimations on the blood pressure of the cat or a fluorescence biochemical method The histamine test makes use of the effect of physiological stimulation of the adrenal medulla by this substance A typical attack provoked by the intravenous injection of a small amount of histamine can be terminated by the administration of phen tolamine (Rogitine)

Presacral air insufflation may lead to a clear delineation of the tumour in tomograms of the renal regions

age of sixty therefore hypertension gives way to atherosclerosis as the dominating aspect of the vascular disorder

At and below this age however diabetes bears a close relation ship to hypertensive vascular disease These disorders are so intimately associated that in the presence of the latter it should be almost obligatory to carry out a glucose tolerance test diabetes will sometimes be found not only to precede but some times to follow some complication such as a coronary thrombosis

important to realize that it may be a precursor of malignant hypertension Patients with this symptom should be examined repeatedly with the ophthalmoscope

Every patient with hypertension should have the fundus oculi examined at intervals varying according to the severity of the hypertension and the symptomatology Normal fundal vessels are a favourable sign but thickening of the arteries with compression of the underlying veins at the points of crossing will indicate complicating arteriolar thickening Retinitis with haemorrhages and exudates is a serious sign and papilloedema puts the case into the malignant class It is at this last stage that essential hypertension becomes complicated by severe renal damage which is very rare in the earlier stages

While early diagnosis is desirable if any preventive measures are to be taken to reduce the severity of complications it must be realized that in many instances the doctor will not be consulted until some alarming complication has made its appearance Such complications may vary from cerebral haemorrhage to permanent or transient strokes (encephalopathic attacks) left ventricular failure or even advancing uraemia Description of these manifestations is outside the scope of this chapter

Atherosclerosis is a common accompaniment of hypertension Evidence that it is promoted by high intravascular pressure includes its rarity in the pulmonary vessels except in the presence of pulmonary hypertension On the other hand it is often independent of hypertension and is found to an increasing degree in all autopsies after the age of forty whatever the cause of death The two disorders may thus be independent but their effects overlap one another Hardening of the large arteries produces a systolic hypertension without affecting the diastolic pressure Narrowing of the peripheral arteries may lead to a total reduction of the vascular bed and thus promote some elevation of the diastolic pressure as well It has been shown in long term studies that after the age of sixty years moderate elevation of the arterial pressure (systolic not exceeding 200 mm mercury) is almost without influence on the longevity of women and only increases the death rate in men by about fifty per cent over that of the population as a whole at this age After the

DYSPNOEIC CARDIAC INSUFFICIENCY

Dyspnoea is an early symptom of congestive heart failure and it is therefore important to recognize its cardiac origin for early congestive heart failure is easier to control than advanced failure Early diagnosis is of special importance when a treatable condition precipitates failure in patients with underlying heart disease for instance the onset of failure may be due to **anaemia** (acute or chronic) to **respiratory infection,** to the onset of an **arrhythmia,** or to the development of **an active cardiac lesion** such as acute rheumatism bacterial endocarditis or a coronary vascular accident Cardiac dyspnoea may also develop in circumstances when failure to treat it promptly may be unusually dangerous as for instance during pregnancy

True dyspnoea was defined by Meakins as consciousness of the need for increased respiratory effort Dyspnoea is of course physiological if sufficient effort is undertaken and the sensation of breathlessness arises when ventilation is somewhat less than the maximum possible ventilatory volume If the maximum ventilatory volume is reduced by disease then effort dyspnoea occurs at a lower level of ventilation True effort dyspnoea is present when comparatively slight exertion evokes the sensation of breathlessness

The complaint of breathlessness does not always mean that true dyspnoea is present for further questioning may reveal that the sensation experienced is not that which occurs near the limit of normal activity but rather a feeling of difficulty in filling the lungs as if it were not possible fully to expand the chest and this discomfort may be relieved when a single sighing respiration is made This type of respiratory discomfort is commonly associated with effort syndrome and is of psychogenic origin It occurs as often in patients with heart disease as in those without a cardiac lesion but it is not in any way due to heart disease Another type of respiratory discomfort which is not true dyspnoea is psychogenic hyperventilation This is often more obvious to the onlooker than to the patient and it is characterized by its variable relation to exertion for a certain amount of effort may evoke breathlessness one day yet the same exertion can be undertaken the following day without any symptoms Nor is this

CHAPTER VI

CARDIAC INSUFFICIENCY

By A Morgan Jones

IN clinical practice it is necessary to know when limitation of physical activity is due to underlying heart disease and to understand how the heart lesion has given rise to this situation it is in this sense that the term cardiac insufficiency is used here Naturally this includes limitation of activity in congestive heart failure which is a well defined clinical entity in this case dyspnoea due to pulmonary congestion is an early symptom But the onset of dyspnoea due to congestive heart failure is not the only way in which heart disease can reduce capacity for effort In some cases the cardiac output is almost fixed and cannot increase on exercise in these circumstances activity is limited by a sense of exhaustion and in a proportion of cases by effort syncope due to a fall of blood pressure on exertion In other cases there is no heart failure in the sense of reduced pumping power of the heart yet there is cardiac insufficiency for the patient s activity is restricted by effort pain due to coronary disease It is in this broad sense that the term cardiac insufficiency is used in this account and this term is not to be regarded as synonymous with congestive heart failure or forward failure or circulatory failure or coronary insufficiency but includes all these syndromes in which for one reason or another the patient s activity is limited by the presence of heart disease This purely empirical approach is chosen because it is practical and useful at the bedside and does not lead to complicated definitions and classifications of heart failure based upon physiological and pathological concepts sometimes as yet imperfectly understood

there is a tendency to get out of bed and go to an open window If the dyspnoea is very severe however most patients get no further than sitting on the side of the bed Such dyspnoea is due to acute pulmonary congestion and is induced by the supine posture during the night Pulmonary congestion is reduced by sitting up and even more by allowing the legs to hang down Haemodynamic studies indicate that lowering the legs allows blood to pool in them and in quantitative terms this may be haemodynamically equivalent to venesection of a pint of blood The relief of paroxysmal dyspnoea by sitting on the side of the bed is therefore easily explicable the patient should never be hurried back into bed and may indeed be encouraged to spend the rest of the night in a comfortable armchair

Sometimes paroxysmal dyspnoea is evoked by emotion This may be very striking in mitral stenosis—as in a patient who was able to play quite strenuous tennis yet developed acute pulmonary congestion at a clinic while lying on a couch awaiting examination In this case it had previously been quite difficult to be sure that her attacks of breathlessness were not of psychogenic origin but the numerous moist sounds present in her chest during the attack left no doubt as to the cause of her dyspnoea If the attack of acute pulmonary congestion progresses it can lead to pulmonary oedema and this will be marked by the development of coarse bubbling moist sounds in the chest increasing cyanosis and the production of frothy sputum which may become copious and pink owing to staining with blood Early recognition of the nature of the attack is important for treatment at once by the correct posture morphia and perhaps intravenous aminophyllin will soon abort an early attack but may not avert a fatal outcome at a later stage Sometimes especially if the patient when first seen is cyanosed and the veins are engorged prompt venesection may be dramatic in its effects It should be emphasized that acute pulmonary oedema may be an early sign of this kind of cardiac insufficiency and may precede any appreciable effort dyspnoea It is particularly liable to occur in this way in severe mitral stenosis in progressive hypertension and of course as an initial symptom in acute myocardial infarction

type of hyperventilation always confined to exertion it may occur for emotional reasons when the patient is seated or even in bed usually before sleep but it sometimes starts after the patient awakens during the night especially if he (or she) is startled or worried

True dyspnoea on effort has of course many causes other than heart disease Obesity is the commonest of these and often develops when the patient has been advised to give up activity perhaps unjustifiably or if illness of cardiac or other origin has compelled confinement to bed In these cases general unfitness is added to the burden of increasing weight. Similarly increasing age and excessive smoking of cigarettes may contribute to this symptom Dyspnoea due to pulmonary disease often complicates heart disease and usually arises from emphysema A history of recurrent bronchitis and chronic cough with sputum together with the shape of the chest and the limited chest expansion will suggest that there is a pulmonary factor in the dyspnoea Numerous other non cardiac causes of dyspnoea such as pul monary fibrosis consolidation collapse pneumothorax or pleural effusion and general causes (for instance severe anaemia) must also be excluded

When it is established that a patient with organic heart disease has true effort dyspnoea and extra cardiac factors can be excluded it remains essential to search for confirmatory evidence that the dyspnoea is cardiac and this can usually be found on examination of the lungs In many cases fine moist sounds will be present at the lung bases but their absence does not exclude a significant degree of pulmonary congestion Radio logical examination of the lung fields gives a much more precise idea of the extent and probable cause of pulmonary congestion than clinical examination alone In the more severe degrees of chronic pulmonary congestion short transverse opacities in the costophrenic angles (Kerley s lines) are often present.

Cardiac dyspnoea is of course not always exertional In hyper tension and aortic valve disease and less often in mitral stenosis paroxysmal dyspnoea may occur This may be quite independent of effort and may awaken the patient in the night The most striking sensation associated with paroxysmal nocturnal dyspnoea (cardiac asthma) is a feeling of suffocation and lack of air and

so inclined that the venous pulsation is maximal in the jugular veins the vertical distance between the highest point of pulsation and the sternal angle is the venous pressure Normally the highest point of pulsation lies one or two cm *below* the sternal angle and the normal range is from 7 cm below to 3 cm above the sternal angle A raised venous pressure is present in conditions other than heart failure such for example as constrictive pericarditis pericardial effusion increased blood volume (as in pregnancy) tricuspid stenosis venous obstruction in the thorax emphysema and raised intrathoracic pressure such as is seen *with a pressure pneumothorax or a large pleural effusion* An important manifestation of a general rise in venous pressure is tender distension of the liver This may give rise to spontaneous pain especially on effort.

Most of the manifestations of congestive heart failure are due to failure of the right ventricle This may occur alone but more often there has been previous failure of the left heart due either to left ventricular failure or to mitral obstruction with consequent raised left auricular pulmonary venous and ultimately pulmonary arterial pressures Left ventricular failure has certain features of its own and these must be appreciated if it is to be recognized before it gives rise to right ventricular failure The triad of left ventricular failure is the combination of *paroxysmal nocturnal dyspnoea* with *presystolic gallop rhythm* (in about two thirds of cases) and *pulsus alternans* which is present in about one third of cases Presystolic gallop rhythm is a most important sign for when typical it is diagnostic It is usually associated with a rather rapid heart rate and the three sounds best heard at or internal to the apex are of similar intensity and nearly equally spaced A double impulse is often palpable Pulsus alternans is usually observed on taking the blood pressure for only alternate sounds come through during the initial five to fifteen cm of decompression this difference of pressure is not easily appreciated by palpation of the radial pulse

In summary therefore the diagnosis of early dyspnoeic cardiac insufficiency depends upon the recognition of true effort dyspnoea and the simultaneous observation of associated features (such as oedema raised venous pressure and an enlarged tender liver in right heart failure and presystolic gallop rhythm and pulsus

Cardiac oedema is not usually associated with early dyspnoeic cardiac insufficiency but its presence does provide confirmation of the cardiac nature of the dyspnoea Nevertheless even the association of dyspnoea with oedema is insufficient to establish a cardiac etiology Both may occur for example in later pregnancy without any heart disease while in older people it is quite common to observe dyspnoea due to emphysema combined with oedema from venous insufficiency in the legs often with a history of old thrombophlebitis and varicose veins It may be difficult to recognize oedema due to local venous causes in the legs for the superficial veins are not always obviously varicose Such oedema is however usually asymmetrical one leg being constantly more swollen and the history of ankle oedema is usually of long duration often preceding the onset of dyspnoea by several years This story is unlike the sequence of events in congestive heart failure for in this condition dyspnoea usually precedes oedema

There are other types of oedema to be differentiated from cardiac oedema During the premenstrual phase as in pregnancy there is a tendency for retention of salt which may lead to ankle oedema especially in women whose work keeps them sitting or standing but otherwise relatively inactive In general women are much more liable than men to oedema other than that of cardiac or renal origin Renal oedema may be unassociated with dyspnoea and tends to affect the face especially below the eyes in the morning However it is not always easy to exclude renal oedema and estimation of the plasma proteins may be necessary in doubtful cases Lymphatic oedema pits very little or not at all on pressure is usually of long duration and is not associated with dyspnoea unless there is another cause for this such as obesity Cardiac oedema is generally recognized when it is present the usual clinical error is to attribute oedema of non cardiac origin to heart disease

An additional feature of importance in establishing the cardiac origin of oedema and dyspnoea is the presence of raised venous pressure The jugular venous pressure is estimated clinically by observing the relationship of the upper level of pulsation in the neck veins to the sternal angle of Louis This reference point is chosen because it is about 5 cm above the right auricle in both supine and sitting positions The patient should be supported with the trunk

disease it is not uncommon to overlook the onset of anaemia during pregnancy and this has obvious dangers and therapeutic implications Often the precipitating factor is intra cardiac such as the onset of an arrhythmia (usually auricular fibrillation) the development of painless ischaemic myocardial damage or the recrudescence of active cardiac rheumatism or subacute bacterial endocarditis If uncontrolled auricular fibrillation active cardiac rheumatism or subacute bacterial endocarditis are present treatment of these complications is obviously essential to success in the control of the heart failure and this is equally true of respiratory infections anaemia and thyrotoxicosis

FIXED OUTPUT CARDIAC INSUFFICIENCY

When congestive heart failure is advanced the cardiac output may become almost fixed at a low level attempted exertion does not then increase the output and may even reduce it This situation is not what is meant here by fixed output cardiac insufficiency for in such instances this is a manifestation of advanced heart failure However in certain severe obstructive lesions of the circulation notably aortic and pulmonary stenosis (without a shunt) and severe pulmonary hypertension (either primary or secondary to tight mitral stenosis) a similar situation may occur in the absence of advanced congestive heart failure In these circumstances the patient complains of inability to undertake exertion owing to a sensation of exhaustion and a feeling of heaviness and uselessness in the limbs If this is not recognized as a cardiac manifestation there is a tendency to attribute it to psychogenic factors The symptom of exhaustion is often difficult to evaluate but when it is of organic origin the patient usually consults his physician about it before it has been present for many weeks On the other hand psychogenic exhaustion has usually been present often intermittently for years before medical advice is sought Also cardiac exhaustion unlike psychogenic exhaustion occurs only on physical exertion and prevents almost all activity

In some cases cardiac exhaustion is associated with effort syncope The way in which exertion may precipitate syncope is illustrated by the following case A man was referred by his

alternans in left heart failure) together with the exclusion of other causes of dyspnoea and the recognition of the presence of heart disease which might lead to dyspnoea The character and severity of the heart disease are of paramount importance for this must be of such a kind and of sufficient severity to make the presence of heart failure likely *It is a not uncommon clinical* error to assume that oedema or dyspnoea or jugular venous congestion are due to heart failure when more critical evaluation would show that only trivial heart disease unlikely to lead to heart failure is present In this connection the size of the heart is of importance for large hearts are liable to fail Naturally heart size is best estimated radiologically but clinical estimation is of considerable value The character of the cardiac impulse is important for a heaving impulse is often the best indication of left ventricular hypertrophy Nor should a right ventricular heave in the centre of the precordium be over looked Unfortunately not all forms of dyspnoeic cardiac in sufficiency are associated with clinically obvious cardiac enlarge ment This is particularly true of some cases of obstructive valvular lesions such as mitral aortic and pulmonary stenosis and of some destructive myocardial lesions especially ischaemic lesions Sometimes the primary cause of the cardiac insufficiency is not very obvious in hypertension the blood pressure may have fallen by the time heart failure makes its appearance perhaps due to ischaemic myocardial damage while in mitral stenosis the typical murmurs may become much less obvious if the cardiac output falls during failure so that the mitral blood flow is much reduced

These considerations are all concerned with the diagnosis of the underlying cause of the heart failure In about fifty per cent of cases however there is *a precipitating factor which is the immediate cause of failure* The recognition of this precipitating factor is of great importance in management for success may depend upon its treatment The immediate cause of failure may be extra cardiac such as the onset of anaemia the development of a respiratory infection or the onset of thyrotoxicosis Symp toms of dyspnoeic cardiac insufficiency often develop for the first time owing to the haemodynamic changes of pregnancy but this will rarely be overlooked In the presence of known heart

has been some recent acute event in the coronary arteries or the myocardium for the therapeutic implications of this situation are *important A recent ischaemic injury to the heart demands that* the myocardium be rested as far as possible for as in other organs good healing is more likely to occur in a resting tissue There is special reason to believe that this is so in the myo cardium for if a zone of ischaemia is created working the ischaemic muscle might well lead to actual infarction If on the other hand adequate time is given for healing by collateral revascularization an adequate blood supply may be restored and painless activity may subsequently be practicable There is now well documented evidence that extensive collateral revascularization does occur in ischaemic areas in the human heart and it is known that the major part of such revasculariza tion occurs within four to six weeks of the initial ischaemic incident Further it has been shown that until revascularization is well developed there is a tendency for ischaemic parts of the myocardium to infarct There is another danger associated with ischaemic zones in the myocardium for sudden death is liable *to occur owing to the onset of ventricular fibrillation or* cardiac stand still though the degree to which this risk can be reduced by rest is hard to assess This account is primarily con cerned with those clinical manifestations which enable us to recognize the likelihood that a recent ischaemic incident has occurred in the heart for it is this which has important thera peutic and prognostic implications

Cardiac pain occurs in three principal ways angina of effort short attacks of pain at rest (angina decubitus) usually relieved by trinitrin and one or more prolonged attacks of severe pain often lasting many hours and usually unrelieved by trinitrin In assessing the likelihood of myocardial infarction the duration of the pain is of great importance If it lasts for hours it is almost certain that myocardial infarction has occurred and this is confirmed if there is shock persistent tachycardia a subsequent low grade pyrexia leucocytosis and a raised sedimentation rate In such cases there is no diagnostic problem If most of these manifestations are absent but the pain lasts for some hours and the patient feels exhausted and ill for a day or two then there is rarely any doubt that a coronary thrombosis has occurred Diffi

physician with the statement that he had been struck by lightning and subsequent examination had revealed a cardiac murmur his physician wished to know whether there was any connection between these It seemed that he had been returning home on foot and when he was about two hundred yards from his house a violent thunderstorm broke out so he took to his heels and ran down the lane He next remembered finding himself lying in a ditch soaked with rain On examination he had typical signs of severe aortic stenosis

Effort syncope is probably due to acute failure of one or other ventricle and it is therefore likely that chronic ventricular failure with congestive signs in lungs or systemic veins will soon follow In the writer s experience however congestive heart failure is not always present when effort syncope first appears Effort syncope is due to inability to increase the cardiac output owing to the obstructive lesion so that when peripheral vasodilatation occurs during exercise the blood pressure must fall These syncopal attacks are dangerous and sudden death is common It is important to appreciate however that in aortic stenosis the low blood pressure predisposes to simple vasomotor syncope this does not occur during effort, but usually while the patient is standing or on sudden change of posture A third type of syncope in aortic stenosis is also unconnected with exertion occurs for no apparent reason and is believed to be due to paroxysmal ventricular fibrillation or standstill

In recognizing fixed output cardiac insufficiency the diagnostic combination is constant limitation of activity by early fatigue and exhaustion with occasional effort syncope Unfortunately unless the cause can be removed or improved this type of cardiac insufficiency is not amenable to treatment and its early diagnosis is of value only in prognosis

ANGINAL CARDIAC INSUFFICIENCY

If exertion is limited by pain due to myocardial ischaemia then in a sense there is cardiac insufficiency This situation may however well be a static one for angina may be persistently evoked by the same amount of exertion for many years Clinically the important thing is to be able to recognize when there

Sometimes however angina decubitus persists in bed and these are dangerous cases Anticoagulant treatment should then be continued until there has been no pain for three weeks even if this means two or three months on anticoagulants In practice it is important to recognize the pattern of events at an early stage and not to delay the initiation of treatment until the pain is ended by a devastating myocardial infarction such as may well occur It is recognized that in at least one quarter of cases of classical coronary thrombosis the typical attack has been pre ceded by symptoms of acute coronary insufficiency

The place of electrocardiography in the early diagnosis of coronary insufficiency has been briefly mentioned but it is essential to have no illusions about its value The history is the important thing and if the electrocardiogram is helpful so much the better But if it does not reveal any diagnostic changes this should not be allowed to create a false sense of security indeed an acute exacerbation of cardiac pain with a normal electro cardiogram is an exceptionally dangerous state of affairs especially if its significance is not appreciated

In assessing the meaning of an acute exacerbation or progres sive increase in cardiac pain in patients with known coronary disease it must be borne in mind that this is not always due to intracardiac causes If the increase in symptoms is slight and gradual then factors such as increase in body weight colder weather or increased physical activities may be sufficient to account for it A more striking increase may be due to the onset of thyrotoxicosis or the development of anaemia If a major gastro intestinal haemorrhage occurs very severe and prolonged cardiac pain may develop and in these cases the electrocardio gram will often show widespread S T depression in leads facing the left ventricle indicative of diffuse sub endocardial ischaemia or infarction In the heart events outside the coronary arteries may lead to the appearance or increase of cardiac pain for in stance the rupture of an aortic cusp or the onset of a rapid arrhythmia such as uncontrolled auricular fibrillation paroxys mal tachycardia or auricular flutter Massive pulmonary embolism or progressively increasing pulmonary hypertension or the rapid onset of aortic stenosis due to aortic valve vegeta tions in bacterial endocarditis are other less common examples

culty begins to arise when there are several short attacks of pain at rest usually lasting less than half an hour or when angina of effort appears abruptly or if previously constant angina begins to occur on significantly less exertion In all these circumstances the essential feature is that there has been an unquestionable change in the manifestations of ischaemic heart disease and the more abruptly this change appears the more likely it is to be significant In some cases the progressive character of the history is striking first the angina starts to increase in frequency and sometimes in severity then short attacks of pain occur almost at rest and eventually even awaken the patient during the night and in some cases there is a final prolonged attack of pain typical of a coronary thrombosis The whole sequence of events usually occurs over a period of two or three weeks but less commonly the progression is slower and may occupy several months This is the type of case to which Master applied the term acute coronary insufficiency It is wisest to regard this term as a purely clinical description and not to assume that any specific pathological change underlies the syndrome (least of all coronary spasm) but there can be no doubt that actual coronary occlusion with myocardial infarction not infrequently occurs in these cases and this can often be demonstrated by serial electrocardiography An electrocardiogram showing only non specific changes or even a normal electrocardiogram does not exclude myocardial infarction for electrocardiograms of this kind are sometimes recorded when there is a typical text book history of coronary thrombosis in which circumstances the history itself would be generally accepted as diagnostic

It is therefore the natural history of the cardiac pain which is the essential criterion of the presence of a recent myocardial ischaemic incident The implications of a sudden or progressive change in the symptoms are important, both therapeutically and in prognosis The mortality within a few weeks or a month or two of such a change in symptoms is quite considerable and comparable with the mortality of the better risk cases of typical coronary thrombosis It is probably at least between five and ten per cent The risks can probably be appreciably reduced by bed rest and a minimum of three weeks anticoagulant therapy provided no pain occurs after the patient is confined to bed

CHAPTER VII

PERIPHERAL VASCULAR DISEASE

BY JOHN H HUNT

THIS chapter deals essentially with the clinical aspects of obstruction arising in the peripheral arteries and veins Some disorders of peripheral blood vessels threaten a patient s life Others change his mode of living by upsetting his activities or his rest, his work or his recreation Even when the circulation in a limb is but slightly affected minor accidents or infections may lead to serious complications Satisfactory treatment largely depends as it so often does *in medicine* on early diagnosis which can usually be made by a family doctor in his surgery or in his patient s home The significant points in the story told by a person with peripheral vascular disease will be discussed in this chapter together with the simple physical examination which is required for correct assessment of the nature of the illness and the stage it has reached

INTERMITTENT ARTERIAL OBSTRUCTION

This condition manifests itself as vasospasm or Raynaud s phenomenon which may be defined as *Intermittent* pallor or cyanosis of the extremities precipitated by cold and aggravated by emotion with the skin a normal colour and temperature between attacks Its recognition is easy the elucidation of its significance may be difficult It is a symptom complex found in many different conditions which occurs without organic peripheral arterial disease or in its presence

Without organic arterial disease, Raynaud s phenomenon is seen in the following circumstances

1 In quite normal people when body temperature is lowered the spasmodic contraction of small peripheral arterioles is then a defence mechanism to conserve body heat. After a long bathe

In practice the principal difficulty in the diagnosis of anginal cardiac insufficiency is to decide whether the pain is in fact due to myocardial ischaemia and this is especially difficult when the pain occurs at rest In these circumstances it is essential to make specific inquiries about previous effort pain for it is surprising how often this is not mentioned spontaneously and how reluctant some patients are to consult a physician about transient pains in the chest which are quickly relieved by standing still especially if they occur only on exertion after meals Yet if such pain has occurred and is similar in character and distribution to subsequent rest pain it is obviously helpful in diagnosis The ensuing rest pain may however be more severe and when this is so it tends to radiate to a wider area so that it may occur for the first time in the lower jaw and teeth or in the arms or hands The site of pain is very variable and may lie anywhere between the mouth and the umbilicus anteriorly or posteriorly and in either or both arms Cardiac pain commonly starts centrally and radiates peripherally but the reverse can occur Its character is described as heavy pressing constricting like a lump aching or burning but not as stabbing or shooting It is in some ways similar to pain from an oesophageal lesion or a hiatus hernia but even if such lesions are present it is wise to be sceptical of their relevance when other features favour a diagnosis of cardiac pain

body is more important than that of the extremities when the body is really warm vasospasm does not occur in the hands or feet however cold they may be when the body is cold colour changes will appear in the extremities even when they are kept relatively warm As the attack progresses pallor cyanosis or both may spread up the hand or foot to the wrist or ankle These colour changes are associated with sensations of tingling numbness burning or tightness sometimes with quite severe pain and with a little swelling of the fingers if the attack has lasted long Recovery which takes place after a few minutes or hours is often preceded by the appearance of red patches on the blue background looking like spots on a plaice Between attacks the colour and temperature of the skin are normal Attacks are more frequent when the weather is cold When the underlying condition becomes worse they may occur also during the summer months the pain that accompanies them is more intense and flakes of ischaemic skin at the tips of the fingers may die and peel off

PERMANENT INCOMPLETE ARTERIAL OBSTRUCTION

The condition of permanent though still only partial arterial insufficiency in a limb gives rise to intermittent claudication It may be recognized from a history of this symptom by seeing persistent colour changes in the extremity and by demonstrating a decrease or absence of its arterial pulsations These three important diagnostic features will be discussed separately They are all much more common in the legs than in the arms The site and rate of the occlusion determine the symptoms and physical signs Owing to the development of a collateral circulation it is a general rule that the longer the obstruction of a main artery takes to develop the less severe will be its effects

Intermittent claudication

This is often the first symptom of partial arterial insufficiency It is variously described by patients as a feeling of intense

in the sea the extremities may become blue and numb with cold

2 In about twenty per cent of healthy people who are known to have a poor circulation or ' Hereditary Cold Fingers In them especially in those who are thin comparatively slight exposure to cold or emotion increases vasomotor tone enough to close their peripheral circulation for short periods This liability often runs in families

3 In true Raynaud s Disease —a severe form of cold fingers and toes without organic arterial disease—which is rare In the writer s opinion only one of Raynaud s original thirty one cases of peripheral gangrene falls into this group It too runs in fam ilies is much more common in women than in men and usually begins between the ages of fifteen and thirty Some local fault in the digital arteries or arterioles almost certainly plays a part in the aetiology of this complaint even though this cannot be demonstrated microscopically The modern view does not support Raynaud s thesis that spasm alone can cause gangrene

4 In cervical rib and other causes of the Thoracic outlet syndrome in which irritation of the cervical sympathetic chain causes vasospasm in the hand on the affected side

5 In cold sensitivity when the digital vessels become tem porarily blocked with agglutinated red blood corpuscles and in which haemoglobinuria may occur

6 In vasospasm from ergot poisoning as in the treatment of severe migraine

With organic arterial disease, Raynaud s phenomenon occurs when normal vasomotor tone from cold or emotion temporarily closes vessels already partially obstructed In this way it appears after trauma to peripheral arteries or when there is scarring around them and in the early stages of any disease of the smaller arteries and arterioles—especially in thromboangiitis obliterans and other conditions likely to cause thrombosis in small arteries in scleroderma and lupus erythematosus in pneumatic hammer disease and other occupational arterial disorders (see Chapter XXV) in polyarteritis nodosa and other rarer conditions

Raynaud s phenomenon from *all* these different causes first shows itself as intermittent colour changes in the hands or feet beginning in the finger tips or toes The temperature of the

Colour changes in the feet

In the early stages of partial arterial insufficiency of gradual onset the feet may be a normal colour Later the toes and dorsum of a foot may become permanently pale bluish or red When there is a marked degree of vasomotor spasm on top of the organic obstruction the development of Raynaud s phenomenon may be the first indication of underlying organic arterial narrowing When the degree of arterial block is considerable lifting the leg makes the foot paler while lowering it turns it a dusky red When the limb has been raised for two minutes—long enough to make it pale—the rate at which this redness appears when it is lowered again can be used as a rough measure of the severity of the arterial obstruction if the foot reddens in fifteen seconds the obstruction is only mild if it takes thirty seconds the block is moderate and if it takes a minute or more a severe degree of arterial obstruction can be diagnosed A difference between the two feet may be important confirmatory evidence

Changes in arterial pulsation in the legs

Partial arterial insufficiency rarely gives rise to symptoms until the pulses are impaired so that diminished or absent pulsations in one of the arteries of a limb are important and consistent findings in this condition They are most easily recognized when the patient is at rest and warm the administration of a vasodilator such as glyceryl trinitrate may occasionally be of help in eliciting this physical sign The dorsalis pedis artery is usually felt most easily on the dorsum of the foot just in front of the ankle along a line extending upwards from the medial side of the middle toe In about one in ten normal people it is absent from the usual position so that being unable to feel it is not always significant The posterior tibial artery is felt just below the medial malleolus in nineteen out of twenty normal persons The popliteal artery is usually palpable in the middle of the popliteal fossa when the patient is lying on his face with leg muscles relaxed and the knee bent to a right angle The femoral artery is felt in the groin below Poupart s ligament with the patient lying flat on his back An oscillometer of the Collins

fatigue a gripping squeezing discomfort ache cramp or pain in their legs brought on by exercise which may make them limp and then force them to stop walking for a while In order of frequency it occurs in the mid calf radiating downwards (soleus claudication) in the foot (pedal claudication) in the thigh from the back of the knee radiating upwards (gastrocnemius claudication) in the front of the shin (anterior tibial claudication) in the hip or in the lower part of the back (gluteal claudication) It generally starts on one side if it is bilateral one leg is usually worse than the other The distance that can be walked before such pain comes on—the patient s claudication distance —may be anything from two miles or more to less than 20 yards depending on the degree of arterial obstruction and the state of the collateral circulation about a quarter of a mile is a common distance The claudication distance of any patient varies with the type of exercise and the conditions under which it is taken walking hurriedly on pavements or uphill is nearly always more troublesome than moving slowly on the level or on grass Three degrees of claudication may be recognized (1) *When the pain comes on with exertion but passes* off as the exercise continues and the patient may be said to walk through his pain the full circulation opening up with exercise rather like getting one s second wind (2) When the pain persists throughout exercise but is not bad enough to stop the patient walking he may then be said to walk with his pain or in spite of it (3) When the pain is bad enough to stop him in his tracks and he finds that he cannot go on He may then develop little face saving tricks—such as gazing into shop windows cleaning his spectacles or turning round as if looking for someone—so that his sudden standing still may not attract undue or unwished for attention Intermittent claudication is often wrongly diagnosed as foot strain muscular rheumatism fibrositis or neuritis In the legs of elderly people these four diagnoses should always be regarded with considerable suspicion until claudication has been excluded Gluteal claudication from obstruction of the iliac artery can easily be mistaken for arthritis of the hip or for sciatica from an intervertebral disc lesion

him a hot poultice is a bad mistake when the condition is really one of incipient gangrene for which heat is the worst possible treatment and for which his leg may have to be amputated in a few days or weeks It is only too easy to operate on an early ischaemic lesion of a toe under the impression that the pain is due to a suppurating corn or bunion or to an ingrowing nail

As the circulation in a limb becomes worse with increasing ischaemia of muscle and nerves intermittent claudication is replaced by *rest pain* this occurs most often in the toes or foot but it may extend further up the leg A persistent, gnawing burning or tingling ache or pain it is generally worse at night and interferes seriously with sleep Sometimes it is felt as sharp stabs like electric shocks up or down the limb It may be aggravated by lifting the leg and is sometimes relieved by lowering the foot over the side of the bed on to a chair Some patients are found rubbing their feet or fanning them with a newspaper for hours on end in an effort to obtain relief With incipient gangrene the pain becomes persistent and agonizing lasting perhaps for days but with little to show for it As it does not respond to ordinary doses of analgesics doctors or nurses may get a cruelly incorrect impression that the patient is fussing unduly With the onset of ulceration and gangrene the pain may be unbearable In diabetics and occasionally in other patients the pain may be less severe perhaps because of associated neuro pathy

Colour changes become more marked as the circulation de teriorates Pallor of the skin after pressure persists for much longer than normal The dusky redness turns to deep cyanosis Later a hard black scab develops with a sharp line of demarca tion between it and living skin Gangrene usually starts at one of the places where ulceration tends to occur in partial arterial insufficiency—at the end of a toe beside a nail or at a pressure point on the side of the foot or on the heel It may extend upwards to involve the foot or lower leg but it rarely reaches above the knee There may be associated thrombophlebitis cellulitis or other infection

The affected skin may be quite cold to the touch numb and with a band of hyperaesthesia above Flaccidity weakness or wasting of muscle may be found Foot drop is not uncommon

type which is as easy to use as a sphygmomanometer, is a valuable instrument for confirming the degree of arterial patency in doubtful cases when palpation of pulses is difficult especially when there is oedema or when the patient is obese The most convenient and useful place on the leg to apply the cuff of this instrument is just below the knee

Other evidence is sometimes helpful in making an early diagnosis of partial arterial insufficiency Temperature changes may make the affected foot and lower leg feel colder than the other side Sensory changes may be found careful examination revealing a little numbness A variety of trophic changes may be detected nail growth may be slow on the affected limb or the nails may be deformed the toes may look shrunken and have no hairs on them and there may be increased liability to paronychia hyperkeratosis or desquamation Small slowly healing ulcers may be present at the end of a toe beside a nail under a toe in the fissure between toes or at pressure points on the outer side of the foot or under it or on the heel About half of such ulcers result from avoidable trauma—cold local antiseptics burns or unskilled chiropody Filling of superficial veins may be slower than normal when the limb is lowered Occasionally a bruit may be heard over the site of a partial arterial obstruction and some way distal to it in patients with intermittent claudication such a murmur can be heard over the femoral artery more often than is generally known In difficult cases arteriography may be needed to establish an early diagnosis

PERMANENT COMPLETE ARTERIAL OBSTRUCTION

Complete organic block of a peripheral artery may occur gradually or suddenly Whether or not gangrene follows will depend on the state of the collateral circulation The longer that an obstruction of a main artery has taken to become complete especially in younger patients the more efficient is the circulation likely to be in the collateral vessels in which there may be however many anatomical variations

Gradual onset of complete block

To tell an elderly patient that his painful toe is just a touch of gout or a little inflammation under the nail and to give

surgical centre for operation, if possible within twelve hours of the onset When all family doctors realize the importance of this many limbs will be saved

When the cause of sudden block is arterial thrombosis one of the predisposing causes of this may be apparent local trauma such as a stab or gunshot wound pressure from a tight bandage plaster or crutch a slowed circulation (as occurs in heart failure post operative shock or any severe illness) or when there is an increased viscosity of the blood and a special tendency to clot Over a period of a few minutes to several hours the patient notices increasing pain coldness or pallor but numbness and paralysis are less common than with emboli Evidence of arterial obstruction may be found on examination The condition may either improve rapidly—as it usually does when pain is not severe and a collateral circulation is already present—or it develops into peripheral gangrene

VENOUS OBSTRUCTION

Blockage of a superficial vein of a limb usually gives rise to no circulatory trouble because the collateral venous circulation here is efficient

Deep venous thrombosis of the main vein of a limb is a more serious matter especially in the leg There may be cyanosis distally with colour changes somewhat resembling those in arterial block especially when the foot is lowered The limb may be swollen and then it is helpful to measure the circumference of the calf every day recording the readings and marking the level of measurement with an indelible pencil The affected leg may be warmer than the other and superficial veins distal to the block may be prominent Secondary varicose veins may develop in a few weeks and may spread up to the abdominal wall Chronic venous insufficiency with gravitational eczema and ulceration may follow

ARTERIOSCLEROSIS

Arteriosclerosis is by far the commonest of all peripheral arterial diseases It usually begins to give trouble between the ages of fifty and seventy but it may do so earlier it is six times

it is due to ischaemia of the anterior muscles of the leg and is one of the manifestations of the so called anterior compartment syndrome Oedema may be present in the foot if the patient has been sitting up all night or has kept his leg hanging out of bed

Sudden Onset of Complete Block

This is usually due either to arterial embolism or thrombosis. If due to embolism the source of the embolus may be obvious or this may have to be carefully looked for auricular fibrillation mitral stenosis recent coronary thrombosis bacterial endocarditis disseminating carcinoma or thrombophlebitis with patent fora men ovale In cases where the source is not found a detached arteriosclerotic plaque may be the cause Arterial emboli nearly always lodge at the bifurcation of a vessel The onset of symptoms is usually sudden there is pain and tenderness which may perhaps only be slight at the point of lodgement with numb ness and tingling of the foot or hand which may be pale and cold A fairly sharp line of temperature change on the skin best detected with the back of the observer s fingers may be found distal to the block The site of this change depends on where the episode has occurred if it is found just above the ankle this suggests that the embolus has lodged at the bifurcation of the popliteal artery if at the junction of the lower and middle third of the calf it indicates a block at the bifurcation of the femoral artery and if between the middle and lower thirds of the thigh it means a lodgement at the bifurcation of the common iliac artery There may be colour changes The superficial veins of the foot and leg may be seen to be empty Arterial pulsation may not be felt distal to the embolus There may be muscular weakness and impaired reflexes and sensation may be markedly diminished The patient may feel ill in himself and tachycardia may be present If the obstruction partially frees itself or the collateral circulation opens up recovery from a peripheral arterial embolus may take place rapidly otherwise gangrene supervenes A great responsibility rests on the general practi tioner to make a prompt diagnosis in cases of arterial embolism so that heparin (as first aid anticoagulant therapy) can be given at once and the patient transferred immediately to a suitable

disease appear in a young man between the ages of twenty and forty who is known to be a heavy smoker It occurs more frequently in the Jewish race than amongst Gentiles It should never be diagnosed in a woman or in a man who is a nonsmoker until every other cause of arterial trouble has been most carefully excluded Smaller arteries are affected than in arteriosclerosis most often those of the feet and lower calves In thrombo angiitis obliterans the hands may sometimes be affected severely this hardly ever happens in arteriosclerosis The femoral artery which is so often involved in arteriosclerosis is seldom badly affected in thrombo angiitis

Symptoms and signs of partial or complete arterial insufficiency are the same as in arteriosclerosis except that, because the arteries involved are smaller it is in the muscles of the feet that intermittent claudication commonly occurs and the distal pulses are more often impaired than the proximal The tips of the toes may burn after exercise and there may be pain and tenderness at the site of the affected vessel Arteriography or arterial biopsy may sometimes be needed to confirm the diagnosis

Veins are affected in about half the cases of thrombo angiitis obliterans usually the small or medium sized veins of the foot ankle lower leg thigh or abdominal wall and generally only two or three inches of each are involved at one time Superficial thrombophlebitis is often the first symptom of the illness it may recur in crops for some years as one form of thrombophlebitis migrans before the arterial element of the disease becomes manifest

Several *episodes of thrombo angiitis obliterans may trouble* a patient over a period of ten or twenty years different segments of the smaller arteries and veins of the limbs being affected on successive occasions or there may be only one or two episodes altogether in the course of a few months Their number and severity are usually diminished when smoking is stopped they are more frequent and severe when smoking is heavy Removal of a septic focus may reduce them In many instances the disease ultimately becomes inactive leaving a damaged arterial tree which is particularly vulnerable to the development of arteriosclerosis in later life

7

more frequent in men than in women Hypertension diabetes gout or polycythaemia predispose to its development Its distribution is often uneven throughout the vascular tree so that it may have already caused symptoms in the eye brain heart or kidneys When the larger arteries of the legs are involved early diagnosis may be impossible before intermittent claudication develops The arms are seldom seriously affected If an adequate collateral circulation is present in the legs the patient s condition may remain stationary for years but usually it becomes progressively worse with periods of remission Sometimes the downhill course is rapid

Examination of the *fundi* may supply useful early confirmatory evidence of generalized arteriosclerosis for the retinal arteries alone are open to direct inspection Palpation of the *brachial and radial arteries* may show them to be tortuous and thickened The *blood pressure* and *urine* should always be tested routinely In some series of arteriosclerotic patients hypertension has been found in thirty five per cent of the cases and glycosuria in twenty per cent Albuminuria may be present A *blood sugar* estimation may help in the diagnosis of diabetes and a *blood urea* estimation in uraemia The *blood cholesterol* is often over 300 milligrams per cent it is normally higher in the elderly than in middle age A *blood count* may show polycythaemia and a *serum uric acid estimation* may confirm the presence of gout. *X ray examination of peripheral vessels* may be helpful in reaching an early diagnosis Irregularly placed patchy dense local deposits of calcium are important they usually appear first in the iliac artery and the proximal half of the femoral artery A fine diffuse deposit of calcium along the line of an artery is of little or no significance it is common in old age and gives rise to no symptoms When the whole arterial tree is calcified (pipe stem Monckeberg s sclerosis) this too is often of little significance as far as arterial blood flow to the limbs is concerned

THROMBO ANGIITIS OBLITERANS

There is little doubt now that thrombo angiitis obliterans is a clinical entity though it is very much rarer than arteriosclerosis One should suspect it when symptoms of peripheral arterial

give rise to a multitude of different symptoms and physical signs —malaise anorexia fever cough haemoptysis pains in the muscles and other symptoms of polymyositis or polyneuritis and loss of weight leucocytosis eosinophilia and a raised erythrocyte sedimentation rate Nephritis with red blood cells in the urine and a raised blood pressure are common findings

When peripheral arteries are involved as they are in only a small proportion of cases small aneurysms may develop as nodules which may be felt in the muscles These aneurysms sometimes rupture causing haematomata in different parts of the body The appearance of any sudden unexplained effusion of blood anywhere in the body should raise a suspicion of this condition Vasospastic phenomena or organic arterial obstruction causing peripheral gangrene may also occur

THROMBOPHLEBITIS

Superficial Thrombophlebitis

Superficial thrombophlebitis may be diagnosed early when pain tenderness redness and a cord like swelling appear along the line of a vein such as the long or short saphenous or one of the superficial veins in an arm Only a segment of a vein is usually involved the thrombus often being more extensive than the inflammatory reaction and the condition subsides after a week or two leaving perhaps a line of slight pigmentation It is a very common condition and may occur after abnormal pressure on a vein—from sitting too long in any sort of chair (especially deck chairs or in a theatre or aeroplane seat watching television or even during a long ride in a car) from tight garters bandages splints or trusses It is found after injury to veins—from a knock against a piece of furniture or when gardening from the trauma of a needle (venepuncture) from a fracture or from chemical trauma (intravenous injection of glucose ethamolin etc)—and also from local infection (cellulitis abscess or a burn) Stasis in varicose veins or in congestive heart failure is a common cause of superficial thrombophlebitis It may be the first symptom of cardiac insufficiency There is an idiopathic

TEMPORAL ARTERITIS

The early diagnosis of temporal arteritis may be made when an elderly person nearly always over sixty years of age slowly or suddenly develops a throbbing headache with tenderness swelling nodularity and loss of pulsation of the superficial temporal or occipital arteries on one or both sides of the head and perhaps with some redness of the overlying skin Wearing a hat may be most uncomfortable There is often associated pain in the face jaw eyes or temporo mandibular joints nearly half the cases develop partial or complete blindness of gradual or sudden onset The eyes may be affected both together or perhaps with an interval of a year or more between Temporal arteritis is more common than is generally realized and it should always be considered when partial or complete loss of vision happens in an elderly person without obvious cause such as cataract or retinal detachment Third cranial nerve palsy with double vision may also occur The local arterial lesions are often preceded or followed by low grade fever muscle pains loss of weight leucocytosis and a raised erythrocyte sedimentation rate This latter is an important finding which is always present at some stage of the illness Sloughing of part of the scalp may occur and pains and ischaemia elsewhere in the body are not uncommon from involvement of other arteries Arterial biopsy may sometimes be needed as confirmatory evidence Early diagnosis of this illness is particularly helpful to the patient because treatment with cortisone may cure him quickly Even without treatment it is usually self limiting ceasing to become worse in from two months to two years Recovery from most of the symptoms may be gradual or rapid but when blindness happens it is usually permanent

POLYARTERITIS NODOSA

Polyarteritis nodosa (or periarteritis nodosa as it is sometimes called) affects many parts of the body Men are more likely to develop it than women and it is commonest in middle life There may be a history of anaphylaxis allergy or sensitivity to sulphonamides or other drugs This hydra headed illness may

angiography etc produces violent thrombotic reactions all over the body affecting even the mesenteric cerebral or coronary arteries with most serious results

On examination the calf muscles when relaxed with the knee flexed and the foot flat on the bed may feel more solid and less flabby—firmer and more like indiarubber—than they normally do Deep vein thrombosis is likely to be associated with some degree of fever and a slightly raised pulse rate sedimentation rate and white cell count Homan s sign (pain in the calf on *forcible dorsiflexion of the foot) is unreliable* Paul Wood has described a test which is useful with a sphygmomanometer band round the thigh pumped up to a pressure of 40 mm of mercury pain is felt in the calf if deep thrombosis is present. Dickson Wright puts a cuff round each calf and pumps them up together tenderness is elicited at a lower pressure on the *affected side Thrombophlebitis in the larger iliofemoral vein* may be associated with pain behind the knee in the lower thigh or buttock and with tenderness over Scarpa s triangle or Hunter s canal Any or all of the symptoms and signs of deep vein obstruction may be present (see p 95)

The sudden and unexpected onset of *pulmonary embolism* may be the first evidence of deep thrombophlebitis A mistake even worse than missing early gangrene is to tell a patient who has an ache in his calf that it is only a little muscular rheumatism which he should shake off by exercise when the condition is really one of deep thrombophlebitis from which a clot, moving to his lungs may kill him the same afternoon Pulmonary embolism of less severe degree is probably far more common in general practice than is usually supposed Many cases are almost certainly missed or wrongly diagnosed as pneumonia pleurisy or coronary thrombosis Any pleural pain in the chest or shoulder occurring during the first three weeks after an operation after the birth of a baby or after a severe injury should be regarded as due to pulmonary embolism until proved to be otherwise It has been said that like lightning the deep thrombosis that *is recognized by the patient does not kill him* In eighty five per cent of these cases this is probably true and the embolism is preceded by neither signs nor symptoms of thrombophlebitis When there *is* evidence of thrombophlebitis in

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type— thrombophlebitis migrans —which does not seem to be connected with any of these other causes sometimes it is associated with high serum uric acid readings with heavy tobacco smoking a persistently low blood pressure or with a septic focus in some there is a hereditary factor others turn out to be early cases of thromboangiitis obliterans

Deep Thrombophlebitis

Deep thrombophlebitis of veins such as the posterior tibial or the sural veins in the soleus muscle may be diagnosed early when tenderness and perhaps cramp like discomfort are felt in the calf after operations and in patients who lie for long periods in bed In the Mayo Clinic it occurred after one per cent of all operations in 1948 After laparotomy it tends to develop when the operation has been lengthy or extensive and particularly when infection or carcinoma is present in the pelvis Further predisposing causes are the lithotomy position dehydration tight abdominal bandaging or strapping with adhesive tape pressure on the calves pillows placed under the knees and pressure from a pregnant uterus Three quarters of the cases of post operative thrombophlebitis are detected between the fourth and fourteenth days after operation in several cases thrombophlebitis appears in another deep vein within ten days In obstetrics thrombo phlebitis is especially likely to develop after difficult forceps deliveries and if there is any degree of puerperal sepsis phlegmasia alba dolens is said to occur after about one in 150 births Deep thrombophlebitis may complicate any severe illness such as pneumonia or typhoid it is especially common in generalized carcinomatosis and may be the first indication of a deep seated growth of the lung stomach or pancreas Trousseau who first drew attention to this diagnosed cancer of his own stomach in this way It may be precipitated by an ab normal stretch or pull on a vein as in axillary thrombophlebitis or other effort thrombosis It happens when the blood is thickened or its coagulability is increased as in dehydration polycythaemia essential *thrombocythaemia (with its great* increase in platelets) and in so called thrombophilia when an injection for intravenous anaesthesia or for pyelography chol

CHAPTER VIII

LIVER DISEASE

By John Richardson

EARLY diagnosis depends on careful analysis of symptoms prompt appreciation of the significance of physical signs and of the results of investigations and consideration of the likely alternatives *The present chapter will follow a similar pattern*

Symptoms which result from diseases of the liver in their early stages are often vague and difficult to interpret at the very time when proper recognition of their significance is of the greatest importance Furthermore when florid symptoms develop the immediate recognition of their origin in liver disease may permit of successful treatment and thus the importance of an early *appreciation of the true nature of the clinical situation* is para mount at all stages of liver disease

When symptoms are vague it is sometimes helpful if they can be grouped under an appellation which of itself suggests the site of the disorder Those imprecise but frequently used terms biliousness or liverishness supply such a grouping Of course people use these terms to describe many disorders entirely unassociated with the liver such as migraine gastritis pyloric stenosis anxiety and depressive states with nausea or even raised intracranial pressure but patients also sometimes employ them when they are suffering from actual liver disease to describe a combination of nausea anorexia flatulence and upper abdominal discomfort Again such symptoms may suggest one of these terms to the clinician and thus lead to early consideration of the possibility of a liver disorder

CLINICAL FEATURES OF EARLY LIVER DISEASE

Nausea which may at first be slight is one of the most frequently encountered early symptoms in liver disorders It is

one calf it may perhaps be that the embolus which reaches the lung comes from the other symptomless leg A clot that has moved to the lung is never found to be organized it has therefore never been firmly fixed to a vein wall Once pain and tenderness in the vessel have developed it is unlikely that a clot will be detached from that site after two days it certainly will not move The value of anticoagulant therapy started immediately lies in the fact that however much phlebitis there may be shown by local pain and tenderness once a patient is on anticoagulants in adequate doses there is little risk of further clotting of the blood with subsequent pulmonary embolism In chemically induced thrombophlebitis as in the treatment of varicose veins with injections of ethamolin pulmonary embolism is extremely rare It is unusual also when varicose veins have clotted spontaneously their openings into the deep veins usually being smaller than the dilated varix in which the clot has formed If the patient is confined to bed however while in this thrombosis mood the clot may spread to deep veins and pulmonary embolism result In the thrombophlebitis of severe illness or from slowing of the blood stream (as in congestive heart failure or generalized carcinomatosis) and in so called idiopathic thrombophlebitis migrans pulmonary embolism is fairly common

Conclusion

In considering diseases of peripheral blood vessels the solution of many an interesting problem still eludes us Why do they develop? Why does arteriosclerosis affect mainly the larger vessels thromboangiitis obliterans the medium sized and scleroderma the smaller arteries? What is the relation between peripheral arteriolar spasm and malignant hypertension? Why does thromboangiitis obliterans appear in men and seldom in women? Why does it affect both arteries and veins while other conditions leave the veins alone? The early diagnosis of peripheral vascular disorders will be much easier when these questions have been answered

which is rarely encountered in the condition from which it must especially be distinguished—cardiac ischaemia

Although an enlarged liver is frequently the first abnormal sign in disorders of the liver a palpable liver is not necessarily an abnormal one especially in persons with a wide costal angle or in emphysema However if the liver edge feels thickened inelastic or hard the liver is abnormal Gross enlargement is most commonly due to secondary deposits of carcinoma and suspicion of their presence will be heightened if the liver feels irregular and knobbly The classical sign of umbilication due to central necrosis of a secondary deposit is frequently demonstrated at autopsy but is only rarely found clinically Chronic parenchymatous hepatitis or cirrhosis from any cause results in a liver which is firm but its nodularity can only rarely be felt on palpation It may appear to be enlarged even when subsequent autopsy shows that it is considerably reduced in size This is thought to be due to forward tipping of the liver from laxity of its ligamentous attachments and the reduced size may be suspected clinically from the absence of dullness on percussion in the axilla

The finding of a palpable spleen in a patient suspected of having liver disease is a useful sign If the patient is jaundiced an enlarged spleen suggests that the jaundice may be either retentive in type due to a haemolytic disorder or far more commonly regurgitant due to hepatitis of some type rather than to extra hepatic block Cirrhosis of the liver is by far the commonest cause of chronic hepato splenomegaly in adult life in a temperate climate whereas in children the underlying cause is usually an extra hepatic block commonly thrombotic in the portal circulation Young people with hepato splenomegaly should therefore always be investigated at a centre where special diagnostic techniques can be employed as this may reveal a situation treatable by surgical intervention Hepato splenomegaly also occurs in chronic myeloid and lymphatic leukaemia and in the reticuloses such as Hodgkin s disease More rarely it is found in amyloid disease in some of the lipoidoses and in certain tropical diseases such as kala azar These are rare and unless the cause is obvious the finding of a palpable spleen should

certainly the initial symptom in the commonest disorder of the liver at present observed in this country—infective or viral hepatitis Nausea may be present for some days before jaundice appears and its persistence should always raise the suspicion that some disorder of the liver is present, such as anicteric hepatitis or more rarely that obstructive jaundice is impending from some extra hepatic cause like carcinoma of the head of the pancreas or a stone in the common bile duct Loss of appetite is rarely absent when nausea from any cause is present and has no specific quality in liver disease There is however on occasion a definite alteration in the sense of taste in addition to the loss of appetite when quite apart from the usual revulsion at the sight and smell of fats a distaste develops for sweet or sugary foods previously much enjoyed This is not an uncommon complaint and the same may occur with tea chocolate alcohol and smoking Discomfort rather than pain in the region of the liver or in the back which may be described as a heavy feeling or a dull ache sometimes precedes the development of any abnormal physical sign other than some tenderness of the liver Although severe pain from the liver is rarely encountered it can occur in viral hepatitis amoebic hepatitis cirrhosis or when single or multiple liver abscesses lead to perihepatitis It is uncommon for a patient even with massive involvement of the liver from malignant metastases to experience much pain unless there is a perihepatitis and the more usual complaint is of a dragging sensation in the region of the liver The same is true of primary neoplasms of liver which are however rarely encountered in this country

A past or present history of biliary colic at once draws attention to the biliary system and thence to the liver Biliary colic is a pain of great intensity which occurs in the region of the gall bladder and may spread round to the back or up to the shoulder tip on the right side The pain steadily increases for half to one hour when it reaches its maximal intensity and this may be maintained for several hours before it gradually subsides The persistence of the pain without waxing or waning distinguishes it from colic arising in the gastro intestinal tract but not from renal colic which is also a persistent pain without great fluctuations Biliary colic may cause the patient to sweat profusely and groan in agony It also leads to restlessness of a degree

detoxicate or excrete them Arterial spiders must be distinguished from the commonly seen Campbell de Morgan s spots (cherry angiomata) which are found on the chest and abdomen and also from other naevi This is simple if the characteristics of the arterial spider are borne in mind and if pressure on the body (central arteriole) with a pin s head or a pencil tip causes blanching of the legs They are frequently associated with scattered thread like dilatations of small vessels in the skin especially of the face Arterial spiders like palmar erythema are seen most frequently in chronic hepato cellular disease such as chronic parenchymatous hepatitis (cirrhosis of the liver) especially when this is due to alcoholism but they may also occur in acute viral hepatitis They may vary in number with the activity of the disease and sometimes disappear entirely with improvement in liver function

The changes in sexual characteristics which may occur in both sexes amount to loss of libido and to reduction in the axillary and pubic hair with genital atrophy In women menstruation may become irregular and scanty and in some a degree of virilism may develop with hirsutes of male distribution In males the testicles may atrophy and become soft, and gynaecomastia occurs This may be either unilateral or bilateral and it must be distinguished from a simple increase in fatty tissue by the finding of nodular glandular tissue which may be tender Gynaecomastia like palmar erythema and spider naevi can occur as a manifestation of thyrotoxicosis

Increased pressure in the portal system will eventually lead to signs of portal hypertension although a period of relative freedom from symptoms may persist for some time after their appearance before the dire complications of ascites haematemesis and liver failure appear When portal hypertension is present, collateral venous channels will develop due either to intra or extra hepatic obstruction to the venous blood flow through the liver These show themselves as dilated vessels on the anterior abdominal wall Usually two or three dilated veins are seen to radiate from the umbilicus and the direction of the blood flow in these veins is radially outwards from the umbilical region unless the situation is complicated by pressure from ascites on the inferior vena cava The latter can lead to partial obstruction

always raise the possibility of portal hypertension whilst the absence of splenomegaly casts doubt on such a diagnosis

Examination of the hands sometimes affords valuable evidence when liver disease is suspected The fingernails may be clubbed in cirrhosis and of whitish appearance in chronic liver disease even in the absence of anaemia More useful evidence is provided by physical signs which may be the result of endocrine disorder secondary to liver disease These include the development of palmar erythema and spider naevi altered sexual activity and loss of body hair in both sexes and testicular atrophy and gynaecomastia in males

The so called liver palms of palmar erythema are warm mottled and bright pink in colour The change of colour is especially noticeable over the thenar and hypothenar eminences and to a lesser extent over the heads of the metacarpals The fingertips may also be bright pink on their dorsal as well as their palmar surfaces When the thenar and hypothenar eminences are examined in detail it will be seen that the mottling is due to local dilatation of small vessels in the corium Palmar erythema is seen quite frequently in pregnant women and in patients with thyrotoxicosis and rheumatoid arthritis as well as in liver disease It is also occasionally found in other conditions such as subacute bacterial endocarditis and in normal people It often occurs in association with arterial spiders but either may be present independently

Arterial spiders or spider naevi are so called because they have a central arteriole usually about the size of a pin s head pressure on which blanches the smaller vessels which radiate out from it in the manner of the legs of a spider The size of the whole spider varies from little more than a pin s head to an area about 0 5 cm in diameter and as it contains arterial blood the colour is bright pink or red They occur over the face neck and front of the chest and are rarely seen below the level of the nipples though they are frequently observed on the forearms and hands Arterial spiders can occur in normal people and are seen in increasing numbers between the second and fifth months of pregnancy They are also occasionally present in patients with rheumatoid arthritis They are thought to be due to excess of circulating oestrogens because of failure of a diseased liver to

s still in the loins but the whole abdomen is grossly protuberant and the umbilicus everted Distended veins are seen on the abdominal wall and percussion reveals dullness in the flanks with the patient recumbent or rising from the pelvis when he is sitting The dullness in the flanks may shift when the patient turns on his side A fluid thrill may be present and organs or tumours may be palpable only when a quick dipping movement is made by the fingers resulting in ballottement of the organ

The occurrence of ascites in portal hypertension is not entirely due to the rise in portal pressure Hepato cellular failure results in reduced synthesis of albumin and consequent lowering of the osmotic pressure of the blood The body s attempt to make up for the fluid lost into the abdominal cavity leads to various compensatory mechanisms of salt retention and thus further increases the ascites It is probable that the most important of these is the secretion by the adrenal cortex of aldosterone a powerful naturally occurring salt retaining hormone increased amounts of which are found in the urine of patients with ascites or oedema from any cause—so called secondary aldosteronism Sodium retention may also be due in part to reduced renal blood flow possibly by increased secretion of the anti diuretic hormone (A D H) from the neuro hypophysis The abnormal retention of salt by these patients dictates the only practical means of treatment namely drastic restriction of sodium intake and attempts to increase its excretion

Ascites is often accompanied by oedema of the ankles and sacrum and distension of the neck veins also due to hypoproteinaemia and salt retention Pleural effusions are not uncommonly found and are more frequent on the right side They are probably due in part to hypoproteinaemia but may result in some degree from raised intra abdominal pressure leading to the passage of fluid from the peritoneal cavity through the diaphragmatic lymphatics into the pleural cavities

JAUNDICE

There is no sign which draws attention to the possible organ of its origin more certainly than does jaundice to the liver and indeed in most instances the liver is primarily involved There are however a number of circumstances in which failure of the liver to

of the systemic drainage from the lower part of the body and consequently to the appearance of distended abdominal veins running upwards from the pubic region and draining into the superior vena cava The presence of a mass of veins radiating from the umbilicus together with a venous hum heard in the region of the xiphisternum indicates that there is obstruction of a para umbilical vein arising from the left branch of the portal vein and thus the block is intra hepatic A full caput medusae is seldom seen Portal hypertension may cause varices to develop where the portal and systemic circulations join at the lower end of the oesophagus and the cardiac portion of the stomach and also at the anus

Haemorrhoids are so common that their presence is of little diagnostic importance The usual situation from which a serious haemorrhage may follow the development of portal systemic collaterals is in the submucous layer of the oesophagus and the cardiac portion of the stomach where the left gastric and short gastric veins anastomose with the oesophageal veins The varices or oesophageal piles which result may rupture with most serious consequences not only from the severity of the blood loss but also because haemorrhage may in itself precipitate the onset of acute liver failure A barium swallow is a safe means of demonstrating the presence of oesophageal and upper gastric varices but results are positive in only about forty per cent of patients Oesophagoscopy and gastroscopy by which the varices are readily demonstrated carry considerable danger from rupture of thin walled vessels and must only be carried out by experts

Patients with ascites due to portal hypertension are usually ill with evidence of wasting and dehydration Ascites may however come on suddenly as a result of gastro intestinal haemorrhage an exacerbation of hepatitis or thrombosis of the portal vein in a patient with hitherto symptomless cirrhosis Under these circumstances the patient may appear relatively well nourished and in reasonably good condition It is in such cases that the detection of ascites may be difficult since the classical signs may be absent and the presence of fluid in the abdominal cavity is only to be inferred from the appearance of fullness in the flanks Of course when the patient is obviously ill and the ascites large its demonstration is simple The maximal swelling

Several points in the history other than the colour of the urine and stools may suggest the cause of the jaundice A story of biliary colic will clearly suggest the presence of gall stones while a period of considerable discomfort lasting possibly for several days with nausea and anorexia is more characteristic of hepa titis than of blockage of the common bile duct Symptoms from a primary growth will make it likely that the jaundice is due to secondary deposits in the liver although it is unusual for this to be severe unless the liver is considerably enlarged Painless relatively symptomless obstructive jaundice in middle aged or elderly persons always raises the fear of carcinoma particularly of the head of the pancreas However none of these common associations is without exception and detailed observation with some investigations will almost always be required in order to reach an early diagnosis

LIVER FAILURE

Some patients with severe hepato cellular failure develop a characteristic foetor hepaticus which has been variously de scribed as corpse like sweetish and faecal or like a pig sty Description is difficult but once this particular odour has been experienced its further recognition will rarely present difficulty The substance which causes the odour has not yet been identified but its intensity varies with the state of the bowels becoming less for example after adequate evacuation in patients with a wide porto caval anastomosis This suggests that the substance passes from the bowel by way of the portal vein to the systemic circulation Foetor hepaticus may be associated clinically with neurological changes and coma states which are also found in the presence of extensive porto caval anastomoses

As with ascites neurological changes in liver disease which are usually of late onset and sinister significance may also on occasion be the first evidence of liver disorder which is presented to the doctor Here there is some difficulty in terminology The designation cholaemia can no longer be accepted since it is not the accumulation of bile acids in the blood which causes these symptoms Again the most widely used connotation namely hepatic coma can hardly be applied to a conscious

excrete bilirubin is of secondary importance and the main disorder is either some form of metabolic disturbance heart disease with congestive cardiac failure or a haemolytic state

The normal liver can excrete no more than three times the normal load of bilirubin Haemolytic or *retention jaundice* thus occurs when a normal liver is presented with an excessive load of bilirubin produced by destruction of red cells through haemo lysis Jaundice of similar type may also occur when the liver already congested as a result of chronic congestive cardiac failure is presented with a comparatively small excess load of bilirubin such as follows haemolysis in a pulmonary infarction

Jaundice far more commonly results however from disorders of the liver— hepato cellular or parenchymal jaundice—or from extra hepatic blockage of the biliary passages— obstruc tive jaundice Both obstructive and hepato-cellular jaundice are known as *regurgitant jaundice* as the bile regurgitates into the blood stream This occurs either from pressure on the extra hepatic bile passages or from disease of the liver cells themselves

The first step therefore in the elucidation of the cause of jaundice in a patient is to determine whether it is retention or regurgitant in type and if the latter whether the lesion is intra or extra hepatic The absence of bile in the urine indicates that the jaundice is acholuric and of retention type The presence of unpigmented pale stools shows that bile is failing to reach the bowel and that the jaundice is regurgitant In this type of jaun dice bile appears in the urine

The reason why bile is found in the urine in hepato cellular and obstructive jaundice but not in retention or haemolytic jaundice cannot be satisfactorily explained until the method of bilirubin excretion is fully known In general however it may be said that in regurgitant jaundice with complete obstruction the stools contain no colouring matter that in hepato cellular jaundice they are pale though some of the stercobilin to which the normal colour of faeces is due comes through to the bowel and that in haemolytic (retention) jaundice they are abnormally dark due to excess stercobilin On the other hand urobilinogen (converted from reabsorbed stercobilinogen) is found in excess in the urine of patients with retention jaundice is absent in obstructive jaundice and varies in hepato cellular jaundice,

be excluded Help may be obtained from a history of factors precipitating the coma and from the presence of foetor hepaticus

The common factors which precipitate hepatic coma are haemorrhage from oesophageal or gastric varices and intercurrent infections Coma may also follow the withdrawal of large amounts of ascitic fluid Alcoholic bouts or excess of protein in the diet may also act as precipitants as may such commonly used drugs as morphia and barbiturates Ammonium chloride and methionine both of which have some place in the treatment of chronic liver disease may precipitate coma by increasing the nitrogen content of the intestines

In the investigation of a case of suspected liver failure the urine must first be tested for all the common abnormalities as well as for urobilinogen and bile since glycosuria may be present in liver disease and should lead also to a search for diabetes mellitus and haemochromatosis The stools should be inspected for colour bulk odour the presence of mucus or blood and tested for occult blood The absence of bile with the presence of excessive amounts of urobilinogen in the urine and of stercobilin in the stools will indicate that the jaundice is of retention type and should lead to a careful haematological investigation to elucidate the nature of the haemolytic process which is present Anaemia is common in liver disease and may be normocytic microcytic or macrocytic There is also a complicated defect of coagulation in liver disorders partially due to failure of prothrombin synthesis in the liver

Estimation of the *serum bilirubin* will give a numerical measure of the degree of jaundice The normal levels are 0 2 1 0 mg per cent jaundice being rarely detectable clinically when levels are below 2 0 mg per cent but obvious at levels of 7 0 or 8 0 mg per cent A *direct Van den Bergh reaction* is indicated by a colour change occurring as soon as Ehrlich s aldehyde reagent is added to the serum Its presence indicates that the pigment possibly a conjugated bilirubin has passed through the liver cells An *indirect Van den Bergh* where alcohol has to be added to the serum and the reagent before a colour develops appears when the pigment which is thought to be bilirubin has not passed through the liver A direct reaction is thus found in obstructive regurgitant jaundice and an indirect one in haemo

patient It is essential that the neurological changes should be recognized for what they are since otherwise the patient will probably be regarded as having a very severe primary mental illness These changes can be temporary or terminal and indeed they frequently herald the onset of coma but they may also be chronic Under such circumstances early recognition of their significance may avert the onset of coma The neurological changes are due to failure of the damaged liver to detoxicate protein products from the bowel and to their shunting into the systemic circulation without passage through the liver

Early neurological changes are difficult to detect since they are concerned with alteration in personality chiefly affecting the emotional reactions and judgement The patient may become unwontedly careless in his appearance and behaviour and be thought to be suffering from a severe psychiatric illness or organic dementia Depression or excessive euphoria may be associated with loss of judgement and accompanied by involuntary movements of the limbs and curious facial grimacing A characteristic tremor may be seen which has been described as flapping since with the hands outstretched there is flexion and extension of the wrists and of the fingers at the metacarpo phalangeal joints The movements come in bursts and may be absent for long periods the rate varying from a rapid tremor to movements separated by several seconds The highly characteristic flapping tremor is rarely seen in other pre comatose states but has been reported in uraemia and in polycythaemic patients with heart failure The legs and face may also be involved in rhythmical movements Such patients often have marked foetor hepaticus All these movements may disappear if the patient passes into a remission but they frequently accompany the increasing lethargy and drowsiness which passes on to coma

Patients in hepatic coma are usually quiet and may appear to be asleep They may however develop convulsions The reflex responses vary considerably but some degree of clonus is often present and extensor plantar responses not infrequently develop as the coma deepens All the commoner causes of coma will require consideration before a diagnosis is reached and particularly in an alcoholic trauma with a subdural haematoma must

stone although only about ten per cent of gall stones have enough calcium in them to render them radio opaque Cholecystography with oral pheniodol or with telepaque is usually profitless in the presence of jaundice due to failure of the liver to concentrate these dyes even with the addition of morphia to cause spasm of the spinchter of Oddi Injection of biligraphin intravenously is also without value as it too is concentrated by the liver but this method is useful in showing up the bile passages after cholecystectomy A barium meal is an essential investigation in chronic jaundice as it may demonstrate pressure obstruction or even invasion of the duodenum as well as revealing an intrinsic lesion of the stomach if one is present while a barium swallow may be useful in demonstrating varices

INFECTIVE HEPATITIS OR ACUTE VIRUS HEPATITIS

Infective hepatitis is the commonest cause of *jaundice and* indeed of liver disease in this country The infection is spread by blood and faeces which contain the virus the symptoms following two to six weeks after contact with the infected material A similar infection transmitted by infected material from transfusions injections of all kinds vaccination or even withdrawal of blood takes longer to develop—about a hundred days —but otherwise does not differ from infective hepatitis It is known as serum or syringe hepatitis The two infections may be caused by a variant of the same virus or by separate viruses

Cases may occur in groups especially in children and in young service personnel but they may be widely scattered without evidence of direct contact with an infected person

The appearance of jaundice is usually preceded by four or five days illness which varies from vague feelings of weariness nausea and loss of appetite to severe nausea vomiting with diarrhoea and total anorexia During this period the patient is *frequently febrile and the temperature may reach 102* or *103* degrees Fahrenheit At this stage there may be a non specific rash which in the presence of *severe headache and neck rigidity* adds the specific fevers mononucleosis and meningitis to the

lytic retention jaundice Hepato cellular (parenchymatous) jaundice is suggested by a *biphasic* reaction in which an immediate colour change deepens on standing

The mechanism of the *sero flocculation tests* of liver function is not properly understood but figures above the normal levels for the tests indicate some degree of hepato cellular damage In parenchymatous jaundice therefore a rise is to be expected whereas little or no increase occurs in obstructive jaundice The *thymol turbidity* is the simplest of these tests and figures above four units are abnormal

The *serum alkaline phosphatase* is raised both in obstructive and in parenchymatous jaundice The reason for this has not been satisfactorily explained but failure of the liver cells to excrete alkaline phosphatase plays a part in its retention in these conditions The normal level in the serum is three to thirteen King Armstrong (K A) units or 1 5 to four Bodansky units per cent The level in the serum rarely reaches as high as thirty King Armstrong units or ten Bodansky units in hepato cellular jaundice whereas these levels are commonly exceeded in obstructive jaundice The figure of thirty K A units is a useful dividing line in clinical practice but there is some overlap and figures lower than thirty K A units are not infrequently found in patients with obstructive jaundice

The combination of a normal thymol turbidity test and an alkaline phosphatase level of over thirty King Armstrong units in a patient with bile in the urine no urobilinogen and pale stools is strong evidence of obstructive jaundice A raised thymol turbidity test with an alkaline phosphatase level of below thirty K A units bile and urobilinogen in the urine and pale but not entirely colourless stools suggests hepato cellular jaundice

The *plasma proteins* may be reduced the reduction affecting the albumin with increase in globulin An electrophoretic strip will show that the gamma globulin fraction is increased The *serum cholesterol* may be much increased in chronic obstructive jaundice from any cause

Radiology

A plain radiograph of the abdomen should be taken in a persistently jaundiced patient as it may show an opaque gall

The reappearance of urobilinogen in the urine is an indication that some recovery is taking place while its final disappearance indicates a good functional recovery of the liver cells The stools will show a varying degree of colour but will not be so pale as when complete obstructive jaundice is present Mention has already been made of leucopenia and of the presence of abnormal monocytes The E S R is usually raised in the pre icteric stage As the jaundice deepens there is a steady rise in the serum bili rubin level and the Van den Bergh reaction is directly positive as it is in the case of extra hepatic obstruction However the colour darkens on standing thus giving the biphasic reaction characteristic of parenchymatous liver damage

Although the total serum proteins are usually normal or nearly so the amount of albumin may be reduced and the gamma globulin increased with consequent production of positive sero flocculation tests These tests however are usually negative in the early stages when they might be expected to be most useful The thymol turbidity test is probably the most widely used normal values lying between 0 4 units The level of alkaline phosphatase is also raised but rarely reaches figures as high as thirty King Armstrong units levels above which if due to liver disease at all are usually due to extra hepatic obstruction The prothrombin time may be raised an observation sometimes of diagnostic assistance in the absence of jaundice

Patients whose illness follows the above pattern usually present little diagnostic difficulty in practice once the jaundice has developed although this is not true of the prodromal period It is however in those patients who do not develop jaundice where real diagnostic difficulty results Their illness may be a very mild one regarded as a simple gastro intestinal infection or a dyspeptic incident Vague symptoms of nausea loss of appetite abdominal distension and upper abdominal discomfort may persist for several weeks the predominating symptoms being inexplicable weariness with persistent nausea a combination which should always suggest the presence of anicteric hepatitis A palpable tender liver with or without splenic enlargement will further draw attention to this possibility and lead to appro priate investigation

gastro intestinal infections which will already have been considered as a cause of the condition The presence of a leucopenia with abnormal monocytes increases rather than detracts from the diagnostic difficulty at this time The onset of actual jaundice not only relieves the patient being usually accompanied by a fall in temperature and an abatement of symptoms but also narrows the differential diagnosis There are however other causes of febrile jaundice which will require consideration including Weil s disease acute ascending cholangitis portal pyaemia amoebic hepatitis and occasionally even such conditions as lobar pneumonia and congestive cardiac failure A toxic cause for the condition must be excluded and at the present time chlorpromazine jaundice is by no means rare Methyl testosterone and thiouracil are also drugs in common use which sometimes lead to toxic jaundice The finding of lupus erythematosus cells (L E cells) in the serum of patients with supposed viral hepatitis raises the possibility of a lupoid hepatitis as a part of systemic lupus erythematosus

Attention may have been drawn to the liver early in the prodromal period by the complaint of discomfort usually a heavy or congested feeling under the right costal margin With the onset of jaundice the liver frequently becomes palpable and is tender without however becoming unduly hard Nor does the edge lose its elastic feel as it slides over the palpating fingers The liver enlargement is accompanied by a palpable spleen in about one fifth of patients and spider naevi with transitory palmar erythema are sometimes seen Lymph nodes in the posterior triangle on the right side of the neck may be enlarged adding further weight to the possibility of the jaundice being due to infective mononucleosis and in the absence of definite jaundice spontaneous bruising or purpura may lead to consideration of a blood dyscrasia There may be pruritus but this is of slight severity compared with that encountered in obstructive jaundice

Bilirubin will appear in the urine in infective hepatitis before a rise in the serum level possibly indicating an alteration in the renal threshold Its presence can be detected by the use of a sensitive test such as Fouchet s reagent or a tablet test Urobilinogen may also be present before the onset of jaundice after which it may disappear as there is little bilirubin in the intestines.

portal cirrhosis Post hepatitic cirrhosis may also develop after an acute lesion has apparently subsided satisfactorily The initial attack has usually been of considerable severity and may have preceded the onset of cirrhosis by several years during which time the patient has been symptom free

The so called post hepatitis syndrome consists of a persistence of symptoms after all evidence of hepatitis has subsided The patients are anxious about their future fail to gain weight and tend to be intolerant of fats They are conscious of their livers and may in fact palpate their own abdomens and find a liver edge This is particularly the case when the patient is himself a doctor Detailed investigation shows that all tests are normal and in these patients a liver aspiration biopsy may be of value not only as a means of making sure that there is no development of cirrhotic changes but also to reassure the patient

PORTAL CIRRHOSIS

Portal cirrhosis sometimes follows virus hepatitis and it is possible that unrecognized mild attacks of anicteric hepatitis may be the cause of some instances of apparently idiopathic portal cirrhosis Alcoholism has long been known to be associated with portal cirrhosis but it is still uncertain whether this results from a direct toxic action by alcohol on the liver cells or from the protein deficiency which is so frequently present in the diet of alcoholics The importance of inadequate protein intake in the production of various forms of liver disease with fibrosis and fatty changes in the liver cells has been shown in those parts of the world where the diet is deficient in protein A history of previous hepatitis alcoholism or prolonged protein deficiency may therefore be of diagnostic significance

Cirrhosis of the liver may lead to few if any symptoms may result in prolonged vague ill health or may threaten or destroy life from the results of portal hypertension or chronic hepatocellular insufficiency—or by a combination of both—with little previous warning

Those patients in whom portal cirrhosis is an incidental finding at autopsy may have had no more than vague dyspeptic symptoms which in an alcoholic were as likely as not due to chronic gastritis

Another circumstance in which infective hepatitis can cause great diagnostic difficulty is when jaundice once developed deepens and persists for four or more weeks In such patients the presence of an extra hepatic obstruction is a very real possibility and definitive diagnosis is of vital importance A palpable spleen would suggest that the jaundice is due to hepatitis while conversely the presence of opaque gall stones in a straight x ray of the abdomen would clearly indicate the probable cause *Examination of the urine and stools and liver function tests* may well prove unhelpful as the findings approximate to those in obstructive jaundice The serum alkaline phosphatase level and the blood cholesterol are often considerably raised in such cases while the sero flocculation tests yield more or less normal results It is in this type of hepatitis that the bile cholangioles are thought to be affected while the liver cells themselves may be relatively spared—a condition to which the term cholangiolitic hepatitis is applied Aspiration biopsy which has little part to play in the routine diagnosis of infective hepatitis may reveal the true state of affairs in these patients but this procedure is not without danger since haemorrhage is common in the presence of jaundice and in most cases laparotomy will have to be performed in order to be quite sure that an extra hepatic obstruction does not exist It is in these patients with a cholangiolitic type of hepatitis that an intravenous infusion of corticotrophin (A C T H) or adrenal steroids has been used in an attempt to distinguish them from those with an extra hepatic obstruction It is claimed that these hormones produce a fall in the serum bilirubin and serum alkaline phosphatase in cholangiolitic hepatitis with subsequent clinical recovery while in extra hepatic obstruction there may be a fall but this is transitory

Patients with infective hepatitis usually recover in six to twelve weeks but some will have a fulminating illness with deepening jaundice foetor hepaticus neurological changes and coma In others the hepatitis will relapse or there may be a recurrence after many months It is important under these circumstances to be sure that some other cause for the jaundice has not occurred such as extra hepatic obstruction

Hepatitis may persist in a subacute form for months either terminating in liver failure or resulting in the development of

temporary recovery of the patient through the restriction of *dietary protein or the withdrawal of ammonium chloride given* to enhance the effect of a mercurial diuretic Again the appreciation that a haematemesis has originated from an oesophageal or gastric varix may lead to a realization that a patient s serious general condition is due to liver failure and thus indicate appropriate treatment the results of which are sometimes so excellent as to amaze the most experienced observers

A clinical picture similar to that of cirrhosis with portal hypertension may occur with diffuse intra abdominal carcinomatosis and in Hodgkin s and allied reticuloses when hepatosplenomegaly occurs with wasting Chronic extra or intra hepatic obstructive jaundice with consequent biliary cirrhosis can resemble portal cirrhosis but the jaundice is deep and portal hypertension appears late

Although congestive cardiac failure with oedema and ascites may be accompanied by a mild or even moderate degree of jaundice this rarely leads in practice to confusion in diagnosis

A rare condition haemochromatosis may resemble portal cirrhosis in certain stages It occurs usually in males and in them there is an inborn error of iron metabolism which leads to deposition of iron pigment in the internal organs The result is diabetes mellitus and cirrhosis of the liver The synonym *bronzed diabetes arises from the curious coloration of the skin* due to an increase in the deposition of melanin The diagnosis may present difficulties but can be established by aspiration biopsy of the liver

CHRONIC OBSTRUCTIVE (REGURGITANT) JAUNDICE OR BILIARY CIRRHOSIS

When a patient is obviously jaundiced the unwary clinician may feel that an accurate diagnosis is near attainment This is very far from the truth and often much care and thought and possibly direct inspection at laparotomy are required before the correct solution is achieved

Regurgitant jaundice due to obstruction may be acute or chronic and the block *either extra or intra hepatic* Pruritus is

Such patients although relatively symptom free may have had some stigmata of chronic liver disease such as palmar erythema or spider naevi or a Dupuytren s contracture which has recently been shown to have a high incidence in patients with alcoholism with or without cirrhosis

The stage of relative freedom from symptoms may persist for some time without cirrhosis being suspected On the other hand evidence of cirrhosis may be sought in order to explain the finding of a palpable firm liver edge with enlargement of the spleen or of a lowered plasma protein with increase in the globulin fraction in a patient with mild chronic oedema of the ankles In such circumstances simple liver function tests will show little impairment with the exception of persistence of urobilinogen in the urine and of an increase of globulin in the serum A more sensitive test such as the ability of the liver to deal with the dye bromsulphalein may be required or a liver *biopsy performed before any significant abnormality is found*

Not infrequently however patients with cirrhosis develop portal hypertension and liver failure as already described They may have muddy complexions due to pigmentation from an excess of melanin and look dehydrated and ill Although the abdomen may become immense the rest of the body tends to waste with loss of muscle bulk as well as subcutaneous tissue If jaundice is present at all it is usually mild and persistent except in the case of some alcoholics in whom it may be deep There may be oedema of the dependent parts and also a marked tendency to haemorrhage with purpura ecchymoses epistaxes and insidious chronic bleeding from oesophageal and gastric varices and piles with the development of severe iron deficiency anaemia Haemorrhages from varices may on the other hand be large sudden and dangerous and frequently precipitate the patient into liver failure with the onset of foetor hepaticus neurological symptoms and hepatic coma

The importance of early diagnosis in cirrhosis of the liver lies in the possibility of withdrawing the responsible toxic substance such as alcohol and in preventing further progress of the condition by giving a diet high in protein On the other hand the appreciation of some of the dramatic manifestations of liver failure such as neurological changes or liver coma may lead to

tenderness over the gall bladder area but the gall bladder itself is usually not palpable when a gall stone is causing the obstruction whereas it can sometimes be felt when neoplastic obstruction of the ampulla is the cause This is the substance of *Courvoisier's law which is however a law with many* exceptions The jaundice may fluctuate with both these common causes of obstruction and this is due in part to variable bile pigment production but the intensity of the discoloration tends to be more constant when carcinoma of the head of the pancreas is the cause It can disappear rapidly when a stone is passed and occasionally this happens when a carcinoma of the ampulla overgrows its blood supply and necroses with temporary relief of obstruction

When obstructive jaundice has been present for a prolonged period the skin becomes a greenish yellow colour due in part to the oxidation of bilirubin to biliverdin Pyrexia suggests that the obstruction is due to gall stones with some cholecystitis and if the fever reaches a considerable height and is associated with rigors there is probably additional cholangitis When the obstruction and cholangitis are intermittent resulting from local oedema of the bile duct often with a stone acting as a ball valve the fever may be intermittent This is the intermittent hepatic fever of Charcot and the occurrence of fever and rigors in a jaundiced patient should always lead to the suspicion that there is a stone at the lower end of the common bile duct Intermittent hepatic fever may also accompany neoplastic encroachment on the ampulla but this is relatively rare

An interesting characteristic of carcinoma of the body of the pancreas is its tendency to be associated with multiple widespread venous thromboses which suggest the diagnosis of thrombo phlebitis migrans These thromboses may be due to the release of trypsin and thus to clotting Whatever the mechanism the presence of thromboses may be useful in raising the suspicion of carcinoma of the pancreas This is always a diagnosis of the greatest difficulty and it will only be made early if the possibility is held constantly *in mind when an elderly or middle aged person* complains of some loss of weight poorly defined ill health upper abdominal discomfort usually worse in bed and relieved by sitting up and sometimes passing through the back These

an important symptom and in its presence the diagnosis of biliary obstruction can sometimes be suspected for a day or so before the occurrence of clinically detectable jaundice but under such circumstances the onset of jaundice is seldom long delayed Chronic retention jaundice due to excessive haemolysis in haemo lytic blood disorders is distinguished from chronic regurgitant jaundice chiefly by the absence of bile in the urine An exception is when a pigment stone in a patient with haemolytic jaundice forms in the common duct and adds an obstructive element to a retention jaundice

Once jaundice can be detected in the conjunctivae and bilirubin is found in the urine investigation must be under taken to establish whether the regurgitant jaundice is due to obstruction of extra or intra hepatic origin

The sudden onset of icterus in younger persons suggests acute virus hepatitis whereas insidious jaundice in the middle aged or elderly raises the possibility of gall stones or of carcinoma of the head of the pancreas or the lower end of the bile passages Gall stones are the commonest cause of obstructive jaundice and their presence should be suspected when previous dyspepsia (which is usually of the flatulent type but sometimes resembles that of peptic ulceration) is associated with periods of upper abdominal pain or actual biliary colic Fair fat forty female and fecund although a hackneyed description is nevertheless a clinically useful aid to memory provided that it is appreciated that none of these features is invariably present

Gall stones occur frequently in men and their presence is by far the commonest cause of chronic obstructive jaundice Although they are usually the cause of some pain before the onset of jaundice they may be quite painless whereas carcinoma of the head of the pancreas which is often painless before jaundice develops may on occasion lead to upper abdominal pain of either constant or intermittent character which not infrequently goes through to the back Carcinoma is usually accompanied by loss of weight which is however not necessarily conspicuous until jaundice and anorexia develop Most patients with gall stones are overweight

The liver may be palpably enlarged in both conditions and tender when obstruction is due to gall stones There is also

to establish the diagnosis beyond doubt Laparotomy should never be delayed for long in regurgitant jaundice It must be accepted as in many instances the only sure way of determining whether a block is due to a stone or a cancer or that the lesion is within the liver itself

Prolonged obstructive jaundice can lead to the slow develop ment of secondary biliary cirrhosis and every care should be taken even if a laparotomy has been done already to ensure that the lesion causing the obstruction is indeed inoperable Such lesions include calculi formed after cholecystectomy post operative strictures of the bile passages congenital lesions such as choledochus cysts and slowly growing neoplasms The diagnosis of biliary cirrhosis should therefore carry with it the possibility of further laparotomy with operative cholangiography The importance of operative cholangiography must be stressed as its use could prevent many instances of chronic obstructive jaundice

Chronic obstructive jaundice often leaves the patient sur prisingly well and deterioration is usually slow The patient is of course jaundiced not only from primary obstruction but from the secondary changes of biliary cirrhosis in the liver These are fibrosis with coalescence of the portal tracts distortion of the intra hepatic biliary passages and contraction of the liver lobules with irregular regeneration of liver cells The liver and spleen are usually sufficiently enlarged to be palpable but the skin provides more useful physical signs It will show jaundice increased pigmentation due to melanin scratch marks as a result of pruritus and possibly xanthomata These appear on the skin around the eyes in the creases around the hinge joints such as knees and elbows and on the buttocks They are yellowish in colour and may be flat or firm and nodular Both types are sometimes seen in the same patient The serum cholesterol has to be raised above 450 mg per cent for at least three months in order for them to develop but levels far in excess of this occur in chronic obstructive jaundice

Absence of bile from the intestines (acholia) may lead through failure of emulsification of fats to mal absorption with fatty diarrhoea and a jejunal deficiency syndrome Loss of protein may be sufficiently severe to result in hypoproteinaemia with

patients may complain of diarrhoea and vague dyspeptic symptoms possibly due to an associated chronic pancreatitis They may also have glycosuria with a diabetic blood sugar curve The importance of making this difficult diagnosis early has been increased by the advances in surgery which make practicable such operations as pancreatico duodenectomy if only the condition has not advanced too far

Little help will be received from biochemical investigation but a barium meal may be of real value Although the appear ances are often normal this may show narrowing of the duodenum in the region of the ampulla alterations in the mucosal pattern due to infiltration or changes in the motility of the duodenum suggesting partial obstruction If the tumour is sufficiently large the stomach may be displaced or the duodenal loop significantly widened Even when the presence of a stone has been excluded and the cause of an obstructive jaundice rightly attributed to blocking by the pancreas at the region of the ampulla the difficulty remains of deciding whether the block is due to a carcinoma of the pancreas or to chronic pancreatitis This latter cause is unfortunately very much the rarer Carcinoma of the head of the pancreas may be associated with secondary interlobular pancreatitis due to blocking of the main pancreatic duct and when this occurs as a primary condition or as a result of obstruction by a stone it may be impossible to distinguish the pancreatitis from carcinoma even at laparotomy and doubt may persist even after expert histological examina tion thus leaving a faint vestige of reasonable hope for the patient.

The presence of metastases will indicate the source of the jaundice unless there is coincidental choledocholithiasis and since secondary deposits in the liver rarely lead to a profound jaundice and glands in the portal fissure even more rarely to jaundice at all a deep colour suggests a block of the ampulla

Another real difficulty which commonly occurs in practice is the differentiation of patients with sub acute hepatitis in whom the jaundice persists for several weeks and where investigations suggest that there is extra rather than intra hepatic obstruction It is in these patients that aspiration biopsy of the liver may prove helpful but in most instances a laparotomy is necessary in order

to establish the diagnosis beyond doubt Laparotomy should never be delayed for long in regurgitant jaundice It must be accepted as in many instances the only sure way of determining whether a block is due to a stone or a cancer or that the lesion is within the liver itself

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oedema and failure of absorption of vitamin D may lead to osteomalacia of vitamin K to bruising or bleeding and of iron to anaemia Biliary cirrhosis differs from portal cirrhosis in its slow progression and in the late development of portal hyper tension and hepato cellular failure Although biliary cirrhosis is usually secondary to extra hepatic obstruction it also occurs as a primary condition with chronic intra hepatic obstructive jaundice This condition is usually known as primary biliary cirrhosis but has also been called Hanot s cirrhosis cholangiolitic biliary cirrhosis (being regarded as a sequel of the cholangiolitic type of infective hepatitis) and xanthomatous biliary cirrhosis. No name is satisfactory and the last is certainly unsuitable as it is now known that the xanthomatous changes are secondary to the biliary obstruction Since the cause of the condition is un known Sherlock prefers to use the term chronic intra hepatic obstructive jaundice Such patients are usually women of early middle age or older who show the usual features of chronic obstructive jaundice including a curious tendency to duodenal ulcer formation They remain remarkably well for long periods but almost inevitably and rightly are subjected to laparotomy as the differentiation of an intra from an extra hepatic obstruc tion is usually otherwise impossible Aspiration biopsy material taken at an early stage may show a pericholangitis and fibrosis characteristic of the condition but the later appearances are sometimes indistinguishable from those following chronic extra hepatic obstruction The whole future of the patient once again depends on accurate diagnosis which will often demand careful visual examination at operation aided by operative cholangio graphy

CHAPTER IX

KIDNEY DISEASE

By Christopher Hardwick

BRIGHT S disease is a term known to patients and doctors alike To both it means a disease of the utmost gravity which leads inevitably to death Although we may still be unable to prevent its fatal outcome there is now much that can be done to retard its progress and to allow the sufferer from chronic renal disease several years of useful life The efficacy of all such measures is greatly improved if they can be applied early This is especially true of infection in the renal tract, when prompt and thorough treatment may lead to the prevention of severe renal damage in later life Such an infection is usually called pyelitis or inflammation of the pelvis of the kidney However it is now known that there is always involvement of the substance of the kidney and so the disease is strictly speaking a pyelonephritis Usually the condition subsides completely but if it continues there will be fibrosis in the kidney causing secondary contraction with severe hypertension and renal failure

Infection of the kidneys is not usually accompanied by any impairment of renal function and in the absence of an anatomical abnormality in the urinary tract response to treatment is excellent In the various forms of nephritis however it is the nephrons themselves which are damaged and renal function is affected The clinical condition which results depends more upon the site of the disease than upon the causal agent Prevention of these disorders may be impossible but their early recognition is essential if they are to be arrested

The early diagnosis of renal disease is facilitated by the frequency with which albuminuria and hypertension occur Both can readily be determined and positive tests for them leave no doubt so that they form a solid basis on which to make a

diagnosis Whilst albuminuria is an invariable accompaniment of renal disease and hypertension is frequently so both may occur with normal kidneys so that some further discussion is necessary

ALBUMINURIA

Cases of renal disease and even renal failure without albuminuria have been described but they are such rarities that when they are recorded in the more erudite medical journals there is always the most lively criticism and discussion For practical purposes such cases may be disregarded and the physician can make a rule that renal disease cannot be diagnosed in the absence of albuminuria Difficulties arise when albuminuria is found without evidence of renal disease as for instance at a routine examination for employment or life assurance

Minute traces of albumin occur in the urine in normal subjects after violent exercise or in the presence of fever Young people who are thin and of poor physique may have a similar leak of albumin after very little exercise—a condition known as orthostatic albuminuria Before such a diagnosis is made strict criteria must be observed The patient must be young and of indifferent physique there should be no history of previous renal disease or of severe scarlet fever or recurrent streptococcal infections The quantity of albumin should be slight and its appearance after exercise or standing must be amply confirmed The centrifuged deposit of the urine should contain no casts or organisms The blood chemistry and renal function must be within normal limits The above tests are commonly carried out in such cases and when the findings are normal a diagnosis of orthostatic albuminuria is thought to be substantiated The importance of pyelography however is not generally recognized and it must be emphasized that a symptomless albuminuria may not infrequently be due to hydronephrosis Judged by these criteria few cases of orthostatic albuminuria are fully investigated It is not so much that harm may be done to the patient by diagnosing benign albuminuria instead of mild chronic nephritis as that it is important not to miss hydronephrosis in the young If this is diagnosed when the kidney is still functioning good results may be obtained by plastic operations on the other hand

if surgical treatment is withheld until the condition is advanced nephrectomy will probably be necessary

HYPERTENSION OF RENAL ORIGIN

It is essential that rigid criteria as to what is and what is not raised blood pressure should be adopted when renal hyper tension is under consideration The diastolic figure is the important one and for adults seen in the surgery or in their homes when not under basal conditions a figure of 100 mm of mercury is taken as the upper limit of normality It must be remembered that a diagnosis of hypertension is not made on the sphygmomanometer readings alone The pulse should be of raised tension and the peripheral vessels thickened while there will probably be cardiac enlargement and almost certainly accentuation of the aortic second sound In addition there may be changes in the vessels in the optic fundi as well as albuminuria

Careful consideration must be given before it is assumed that a patient s symptoms are due to a raised blood pressure Every practitioner is familiar with patients who on routine examina tion are found to have diastolic figures of 140 mm Hg or even higher yet who admit to no symptoms Such cases should be remembered and they should make one hesitate before ascribing symptoms to hypertension when the diastolic reading is only 105 or 100 mm Hg—or even lower The harm that may be done by telling a patient that he has a raised blood pressure needs no further emphasis

Added point to the association of hypertension with renal disease has been given by the demonstration that damage to one kidney may cause a rise in blood pressure which will fall to normal when that kidney is removed In the enthusiasm which followed this discovery it was advocated that all cases of hyper tension should have an excretion pyelogram In consequence many kidneys were sacrificed unnecessarily as a result of in different radiological techniques and dubious interpretations

With the passage of time it has become possible to view this problem in its right perspective and the indications for the investigation and surgical treatment of a unilateral renal lesion

can now be precisely stated In the first place it is recognized that such a lesion is a very rare cause of hypertension and that pyelography is not required in all cases of raised blood pressure Secondly the removal of such an organ is likely to be of benefit only if the hypertension is of relatively recent onset When the blood pressure has been raised for a long time secondary changes occur in the circulation the arteries and arterioles become thickened and irreversible lesions may occur in the primarily uninvolved kidney Under such circumstances removal of a damaged kidney may be wise on urological grounds but a beneficial effect on the hypertension is not to be anticipated It is therefore in young people that these lesions should be sought. All genuinely hypertensive subjects below the age of forty years without obvious pathology require pyelography If such a unilateral renal lesion is found in a young person or in an older patient in whom it is known for certain that the hyper tension is of recent origin then nephrectomy is probably wise This is so even though the patient s condition may be precarious owing to malignant hypertension or even uraemia In such cases a desperate condition calls for desperate measures—and these are sometimes strikingly successful

INFECTIONS

Infection in the renal tract may occur at any age and is more common in females than in males It is one of the causes of fever in the new born and is frequently responsible for much ill health and misery in the aged The causal organism is usually the *Bacillus coli* but *staphylococci streptococci* and *Proteus vulgaris* may be responsible A mixed infection with two or more kinds of bacteria is particularly liable to occur when there is some gross abnormality in the anatomy of the renal tract In a few cases *M tuberculosis* may cause serious disease in kidneys or bladder

The organisms usually gain access to the urinary tract from the bloodstream In females the urethra is a frequent portal of entry Any deformity in the urinary tract causing stasis of the urine will predispose to infection Such lesions include hydro nephrosis or polycystic disease of the kidneys dilatation or kink

ing of the ureters diverticula or trabeculation of the bladder enlargement of the prostate and valves or strictures in the urethra Women during pregnancy are particularly prone to such infections for then the ureters become greatly increased in size and in the later months they are pressed upon by the gravid uterus

The infection may be acute when symptoms begin abruptly and are obviously related to the urinary tract In chronic infections however the symptoms may be more vague and their origin in the urinary tract easily overlooked Thus the patient may complain of bouts of fever of persistent backache or of anaemia and the urinary involvement may be discovered only by direct questioning or investigation

Frequency and pain on micturition are the commonest symptoms The former may be extreme and is usually only slightly less by night than by day In some cases pain may be so intense that the urine is only passed in drops a condition known as strangury The pain is sharp or boring in character and is usually felt in the suprapubic region but may also be in the urethra or perineum There may be pain in one or other loin

Haematuria occurs more commonly than is usually supposed It may be profuse when it is associated with a haemorrhagic condition of the bladder Though pain is usually present in such cases it may be only slight and the distinction from acute nephritis difficult

Fever is usual Most commonly it is moderate in height reaching 101 or 102 F It may be higher and this is one of the conditions in which a rigor may occur Such an occurrence is therefore an important diagnostic point

In the new born acute pyelonephritis may cause fever and dehydration but both these features are easily recognizable and diagnosis should present no difficulty It is in toddlers of the nappy age and a few years after that these problems arise The child may rapidly become very ill with high fever vomiting or even a convulsion When the infection is less acute the child becomes fractious and cries a great deal although the relation of this to micturition may not be realized by its parents Diarrhoea and vomiting may suggest a gastro enteritis and if there appears to be abdominal pain the infant drawing up its legs

when it cries then acute appendicitis or intussusception may be simulated

In chronic cases in adults symptoms referable to the urinary tract may be so slight that the infection may escape detection This may be particularly so in elderly women who may tolerate frequency to a surprising degree thinking it to be a usual accompaniment of advancing years in those who have had children Such patients may seek advice on account of chronic ill health Years of out of bed nights may result in exhaustion and loss of weight Exacerbation of the disease may lead to unexplained bouts of fever Long continued infection may lead to a microcytic anaemia which resists all efforts to correct it

Clinical examination is important less because of positive findings than for the exclusion of disease in other systems This is particularly so in children Despite the sometimes alarming abruptness and severity of the disease in such patients careful clinical examination reveals no tonsillitis or otitis media the lungs are clear and there are no signs of meningeal irritation There may be tenderness in one or other of the renal angles or in the suprapubic region

Whatever may be the age of the patient or his symptoms examination of the urine will always reveal the correct diagnosis A great deal can be learnt from an ordinary specimen It will be acid to litmus and will have an offensive smell like that of rotten fish Albumin may be present When the glass is held up to the light the urine will not be clear but it will have a fine shimmer or sheen Examination of the centrifuged deposit will show cellular debris and motile organisms The exact nature of these bacteria is only to be learnt by culture of a clean or catheter specimen and their sensitivity to the various antibiotics can be similarly determined This may mean the delay of a day or two before treatment can be started but it is necessary in cases which do not respond to simple measures such as treatment with sulphonamides or which relapse soon after such treatment In some patients there may have to be a special search for tubercle bacilli

As has been indicated a structural abnormality in the renal tract may predispose to pyelonephritis Such a lesion can be shown only by excretion or retrograde pyelography Every case

must be considered against this background and the need for radiography or cystoscopy assessed In females there are several portals whereby infection may enter so that a single attack which responds rapidly and completely to treatment can usually be disregarded But delayed recovery early relapse or repeated attacks demand full investigation In males there is a much greater likelihood that a urinary infection indicates an anatomical abnormality and the younger the patient the more likely is this to be so It is clearly impracticable to carry out an intravenous pyelogram in every small child so affected One solution of this problem is to treat these patients not with antibiotics but with adequate doses of potassium citrate which is effective in all straightforward cases Those who do not respond can be given *antibiotics and these selected patients can then be fully investi* gated

Urinary infection is only one way in which abnormalities of the renal tract declare themselves Backache is another very common symptom and this is especially so when there is a hydro nephrosis Though the pain is usually situated in one or other loin *it may be in the midline and in many cases the urine is* normal In these days when a prolapsed disc is such a fashion able diagnosis this less common cause of backache should not be forgotten

NEPHRITIS

The term nephritis is used not only to denote the various forms of glomerulo tubular nephritis but as a generic term to mean any form of renal disease in which there is damage to the nephrons and interference with their function

Four main types of nephritis will be considered—acute renal failure acute glomerulo tubular nephritis the nephrotic syn drome *and chronic renal failure*

Acute renal failure may follow any condition in which there is a state of shock with low blood pressure It is most common after severe injuries and it came into prominence during the Hitler war when it was known as the crush syndrome or as traumatic anuria or uraemia It may occur *after a variety of* other conditions and is to be anticipated after severe burns

major surgery or mismatched blood transfusion or in cases of sulphonamide sensitivity In all these instances the lower regions of the tubules degenerate and may even rupture A condition which is clinically similar but in which there is damage not to localized parts of the nephrons but to a whole area of the cortex of the kidneys occurs in certain obstetrical emergencies Known as acute cortical necrosis of the kidneys it may follow septic abortions or concealed antepartum haemorrhage

In both traumatic uraemia and acute cortical necrosis the symptoms are the same Diminished secretion of urine leading to anuria is the first abnormality and in any patient at risk a careful daily measurement of the urine must be made Normally fifty ounces of urine are passed each day in this disorder the output may drop suddenly or more usually it decreases gradually over a period of three or four days It is usually highly pigmented but of low specific gravity Albumin may be present and analysis shows a low content of urea For a time the patient may seem curiously unaffected by the complete cessation of the function of one of the most important organs of the body Gradually however he becomes listless loses his appetite and begins to vomit He becomes dehydrated the breath is increasingly uriniferous the skin may be covered with a fine white powder of uraemic snow muscular twitching may occur and increasing drowsiness eventually leads to unconsciousness

Investigations in the laboratory will show acidosis with an enormously high concentration of urea in the blood These findings coupled with the anuria may suggest treatment with massive infusions or with hypertonic solutions in an endeavour to stimulate the kidneys to work Such temptations must be resisted for they can only aggravate the condition and hasten the death of the patient The kidneys must be rested and only sufficient fluid given to allow for that lost by respiration and perspiration

Acute glomerulo tubular nephritis is becoming increasingly uncommon Children are usually affected and the attack follows an infection by a haemolytic streptococcus Usually this is an acute tonsillitis and is easily recognized Sometimes however the patient admits to no more than having had a cold whilst in

others the infection may be so mild as to escape detection completely

The onset of the renal disease is gradual the patient feels unwell headaches become troublesome and backache occurs There is a low fever and the patient may vomit Haematuria and generalized oedema develop and examination will show a raised blood pressure whilst laboratory investigation shows that there is nitrogen retention

The haematuria is usually only slight and may not be remarked *upon by the patient Unless it is carefully examined the urine* which is always reduced in volume may only be thought to be concentrated—as would be expected in a febrile patient who is vomiting When seen in a specimen glass against the light however the opalescent or smoky appearance becomes apparent Chemical tests confirm the presence of blood and microscopy reveals red cells in the sediment At times the urine may be bright red and a lesion such as a papilloma of the bladder may be suspected There is however a general constitutional upset and biochemical changes Curiously enough although profuse haematuria would suggest more serious and extensive renal damage this is not necessarily the case and the prognosis in these cases of so called acute haemorrhagic nephritis is usually excellent

The oedema in acute nephritis is never severe and seldom prolonged Swelling of the ankles in the evening is an early feature but in children puffiness of the face in the mornings may be the first abnormality to be noticed by the parents There is an increase in the total amount of body water and the distribution of the oedema is governed by the position of the patient Usually these patients are able to lie flat and so the oedema is distributed generally Occasionally however especially in elderly subjects the increase in the circulating blood volume precipitates an attack of left ventricular failure In these cases the patient is unable to lie flat and so the oedema collects in the most dependent parts with the same distribution as in congestive cardiac failure

The nephrotic syndrome is caused by a variety of conditions These include subacute glomerulo tubular nephritis chronic pyelonephritis renal vein thrombosis amyloid disease myeloma

tosis and systemic lupus erythematosus It is occasionally seen in diabetics and after taking certain drugs such as Troxidone (Tridione)

Oedema which steadily increases is the cardinal feature of this disorder Again swelling of the ankles in the evenings and of the face after a night s rest are its earliest manifestations Gradually the swelling becomes generalized and pleural effusions or ascites may develop so that the patient is considerably in capacitated and becomes so breathless that he cannot get about

Examination shows a pale oedematous individual There is a true anaemia with pale mucous membranes The swelling which pits on pressure is not confined to the lower limbs but especially in patients who have been resting in bed there is a large sacral pad of oedema The chest may show dullness with absent air entry at one or other base indicating a pleural effusion Shifting dullness will be present if there is ascites Apart from some increase in blood pressure the cardiovascular system usually shows no abnormality but in severe cases there may be fluid in the pericardial sac and the cardiac embarrassment which this causes may further contribute to the oedematous state There may also be clinical evidence of some underlying disorder such as amyloid disease or systemic lupus erythematosus

The urine gives the clue to the nature of the complaint It is essential that it should be tested for the presence of albumin in every case of swelling of the ankles however trivial it may seem to be Even on a hot summer s day when a patient whose job entails a lot of standing comes complaining of swollen ankles and a diagnosis of physiological oedema seems obvious a speci men of urine must be examined In nephrosis albuminuria is massive and at times may reach the theoretical maximum of five per cent Microscopy of the urinary deposit shows waxy and hyaline casts

Laboratory studies show characteristic changes From the earliest stage there is a great reduction in the plasma proteins The albumin fraction in particular is reduced and may amount to only one gramme The globulin content is unaltered or may be increased The blood cholesterol is greatly increased The blood urea is usually raised but may be normal

Simple tests of renal function reveal an inability to concentrate the urine after a protein meal or to produce a copious diuresis after a large draught of water The urea clearance will be diminished

In **chronic renal failure** more and more of the nephrons are damaged until there comes a time when they are insufficient to allow normal renal function to continue and renal failure occurs This state of affairs may be brought about by a variety of diseases—as for example glomerulo tubular nephritis pyelo nephritis polycystic disease

Although disease of the kidneys usually runs an inexorably downward course this does not always proceed at a uniform rate For a time it may seem to halt in its progress and there is no deterioration in the patient s condition which may even seem to improve Then however the disease lights up the kidneys are further damaged and the patient becomes more ill Finally there comes a time when the body chemistry is so upset that death ensues

The excretion of waste products by the kidneys is only part of their work It occurs in the course of their more important function of maintaining an even biochemical climate in the body Some idea of the magnitude of this task may be given by considering the fate of the glomerular filtrate The volume of this amounts to 180 litres per day Of this one and a half litres are excreted as urine but all the rest is restored to the circulation When the kidney fails therefore changes will be found in the urine but they will occur to a much greater extent in the bio chemical content of the blood There will be alterations in the concentration of its various constituents which in themselves may give rise to symptoms In addition a microcytic anaemia which is refractory to all forms of treatment is a very constant feature

There is one other important component of the clinical picture Most disorders of the kidney which are present for a long time are associated with a rise in blood pressure Hypertension itself can produce renal damage and in this way a vicious circle is set up nephritis causing hypertension and the hypertension causing further renal damage In some subjects the effects of raised blood pressure may be more severe than those of the renal

failure and such patients may die from strokes or from heart failure with their renal disease only moderately advanced Usually however there is a mixture of biochemical upset severe hypertension and cardiac failure and anaemia

The symptoms of chronic renal disease may appear to be unconnected with the kidneys The patient does not seek medical advice because he has noticed that his urinary output is diminished or because of pain in the renal area He goes to his doctor on account of tiredness and lack of energy and perhaps difficulty in concentrating on his work symptoms which are common and which may easily be dismissed as functional Breathlessness on exertion is a frequent complaint when the anaemia is severe Nausea with a brassy taste in the mouth loss of appetite and vomiting may be troublesome Bad headaches bursting or pulsating in character may affect other patients and a rapid deterioration in vision is not uncommon Bouts of nocturnal asthma will occur in those who develop left ventricular failure while the convulsive seizures of hypertensive encephalopathy so-called uraemic convulsions may demand urgent medical attention

When symptoms are severe or some calamity such as a convulsion has occurred it is usually not difficult to arrive at the correct diagnosis It is when the complaints are vague or seemingly trivial that the diagnosis is likely to be missed This is especially so when the patient is well known to his practitioner and when the anxieties of the man s job or the difficulties in his family life may suggest an easy explanation for the tiredness weakness and general ill health of which he complains On direct questioning nocturnal frequency may be admitted and slight enlargement of the prostate on rectal examination may again suggest a simple diagnosis Full clinical examination is essential however obvious the diagnosis may seem to be and this examination must include careful examination of the urine for albumin The recent introduction of a test for albumin in which a small stick of cardboard is merely dipped into or wet with the urine has done away with the need for test tubes and spirit lamps or b sen burners The test can now be carried out on any patient ith great accuracy

Patients v al ailure are anaemic but they do not have lexion of a simple iron

deficiency anaemia They look ill and are pale and sallow with dry furred tongues and a foetor oris which may be frankly urinferous Oedema is usually absent even a patient with the nephrotic syndrome losing his oedema when renal failure sets in The radial arteries are thickened the pulse full and hard to compress and its rate usually increased The blood pressure is raised and the diastolic figure may be as high as 140 or even 160 mm Hg The heart is enlarged the aortic second sound accentuated and there may be an apical systolic murmur or even a gallop rhythm There may be moist sounds at the lung bases The abdominal wall is usually lax with dry inelastic skin Hard scybalae may be felt in the colon for constipation is a constant feature The kidneys cannot be felt as a rule although both will be palpable if polycystic disease is present The tendon reflexes are usually exaggerated and other signs of neuromuscular irrita bility may be present The fundi may show only hypertensive changes such as narrow and irregular arteries with nipping of the veins in severe cases however exudates flame shaped haemorrhages and papilloedema may be seen—signs of the gravest ill omen

The importance of urinalysis has been stressed Laboratory investigations are required not so much for diagnostic purposes as for assessment of the severity of the case and as a guide to treatment and prognosis An estimation of the blood urea will show at once whether the disease is compensated or whether there is a serious accumulation of waste products in the blood Estimations of the electrolytes may reveal a need for treatment with specific substances in order to combat a particular deficiency such as a very low alkali reserve

Tests of renal function are of value in prognosis Some indica tion of the state of affairs may be obtained by learning that the patient has to get up three or four times at night to pass water More precise information will be given by the dilution and concentration tests and by the urea clearance test In inter preting the results it must be remembered that it is not only the degree but also the *rate* at which the impairment is produced which is important Rapid reduction in renal function is of far graver significance than a slow deterioration over several years

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Patients with chronic renal failure are anaemic but they do not have the pink and white complexion of a simple iron

It is reasonable to enquire to what proportion of patients diagnosed it will be possible to apply this form of surgical treatment The proportion who can be treated by pneumonectomy is small. It is unusual in the many published series of patients treated in this way to find that more than twenty per cent of those diagnosed had resections carried out In the remainder operative treatment is impossible for various reasons a high proportion have intrathoracic or extrathoracic metastases the tumour may be shown on bronchoscopy to be in such a position that it cannot be removed and a number cannot be treated operatively because of deficient respiratory function very poor general condition or other serious disease

In view of the small proportion suitable for surgery methods of treatment which may be offered to those for whom radical operations are not possible are of great importance These non surgical methods are X ray treatment and possibly chemotherapy The patients treated by these methods are on the whole those in whom an early diagnosis has not been achieved Chemotherapy is still in the experimental phase and the results obtained with it are probably not yet of serious significance nevertheless it is probably with this form of treatment that hope for the future lies The results of X ray treatment vary somewhat and depend in part on the selection of patients The best results are obtained in patients without obvious metastases who are in general those in whom surgical excision is also possible For cases with extra thoracic metastases and in poor general condition palliative X ray treatment is usually the only possibility This does not much affect the length of time for which the patients survive but may produce some symptomatic improvement haemoptysis is usually relieved although the symptom sometimes returns cough is often lessened and sometimes completely relieved superior vena caval obstruction is commonly and dyspnoea sometimes relieved Even in this group palliative treatment offers more chance if the condition is not too far advanced when bronchial occlusion has occurred with infection of the lung behind the block X ray treatment is apt to cause serious deterioration although this disadvantage may be partly overcome by the use of penicillin and postural drainage if the bronchus is not completely occluded Thus although any survey of the results

CHAPTER X

BRONCHIAL CARCINOMA

By Howard Nicholson

BRONCHIAL carcinoma is now the commonest carcinoma occurring in men In 1956 18 000 people died of this disease in England and Wales and this number increases year by year Effective treatment is possible in only a small proportion of cases diagnosed and it is worth considering what advantage treatment offers and whether early diagnosis improves chances of survival

Surgical removal of the affected lung offers the best hope of long survival Operative mortality is now in general less than ten per cent and of those patients who survive the operation approximately two thirds live one year almost a half two years and a third five years The relation of these survival rates to the duration of symptoms is not straightforward It would seem reasonable to expect that the longer the duration of symptoms the worse the chance of survival In fact this is true only for patients who have had symptoms for eight months or less There is no doubt that if the growth can be diagnosed within three months of the development of symptoms the chances of survival after operation are at their highest. When the diagnosis is delayed for six to eight months from the beginning of symptoms the survival rate declines Curiously enough when symptoms have been present for longer periods than this the chances of survival appear to improve However this cannot be regarded as a reason or excuse for delay in diagnosis It is really an expression of the differing degrees of malignancy of the tumours removed Those who are still fit enough for operation and who have no overt metastases more than eight months after symptoms referable to the tumour first appear tend to have slow growing localized carcinomas As there is no way of knowing beforehand which growths belong to this group it is clearly wiser to aim at early diagnosis and treatment

tion of the chest It should be remembered that pain down an arm especially the medial border may be caused by involvement of nerve roots by an apical carcinoma of the lung and in a middle aged or elderly man persistent pain of this sort calls for a chest radiograph However the most difficult problem in relation to pain arises in those patients who complain of the more vague and indefinite forms of chest wall pain These pains are often associated with some tenderness of the muscles and apart from this no abnormal signs are found over the chest There is no doubt that bronchial carcinoma is found in patients who present no more than this sort of symptom The problem is to select the few who have serious pulmonary disease from the majority who do not Some patients in whom the pain is transient or clearly associated with some unaccustomed muscular exertion and ceases as the patient becomes used to or abandons the particular effort need not be further investigated For the remainder there is no simple solution and when this kind of pain persists for two to three weeks it is advisable to have a chest radiograph taken

Haemoptysis is a symptom which almost always alarms a patient sufficiently to make him seek advice and it should always lead to full investigation Frank haemoptysis is less common than blood staining or streaking of the sputum and patients sometimes make light of slight streaking or may fail to inform the doctor about it unless directly questioned Continual streaking of the sputum with blood is a most important symptom however slight and if it occurs daily it is an almost diagnostic feature In the presence of haemoptysis even a normal chest radiograph cannot be accepted as precluding the presence of a bronchial carcinoma without further investigation this is referred to later in relation to the special methods of investigation and their limitations

Symptoms which are related to *infection of the lung* usually follow obstruction of a lobar or segmental bronchus The area of lung supplied by the bronchus collapses and infection sometimes leading to suppuration frequently occurs in the collapsed lung The illness resulting from this infection may be regarded as being no more than an exacerbation of the patient s usual chronic bronchitis or if more severe it may be thought to be an

of treatment of bronchial carcinoma is bound to be gloomy at the present time it cannot be seriously doubted that real advantages come from early diagnosis

SYMPTOMS IN RELATION TO EARLY DIAGNOSIS

In this condition as in pulmonary tuberculosis the discovery of abnormal physical signs on examination of the chest makes it likely that the disease is already far advanced Early diagnosis depends in the first place on the recognition that even though abnormal signs are not discovered patients and especially men over the age of forty who present certain symptoms must have their chests radiographed Although radiography has its own limitations which will be considered later by its means reasonably early recognition is possible in the majority of patients The first difficulty arises in relation to the commonest symptom of the disease namely *cough* This frequently occurs at all ages and is especially common in middle aged smokers in whom bronchial carcinoma is most likely to be found A useful rule is that any middle aged or elderly patient whose usual cough becomes worse or alters in any way should be suspected of having a carcinoma and should be radiographed Also if such a patient who has not previously had a cough develops one and it persists for more than three weeks he should similarly be investigated

The next most common symptom is *pain* This may arise in various ways When a carcinoma blocks a bronchus a pneumonia or infected atelectasis may develop in the lung distal to the block The pleura over the involved lung may be inflamed and typical pleuritic pain arises often followed by effusion Secondary deposits of growth in the pleura commonly result in effusion also but not so frequently in pain although this may occur before the fluid accumulates Extension of the growth to the chest wall may produce very severe pain by involvement of nerves and ribs as may secondary deposits in ribs or other chest wall structures In all these circumstances the illness of the patient and the considerable abnormalities found on clinical examination will point to the urgent need for further investiga

formation by the introduction of radio active gold into the pleural space

The symptoms that arise from *distant metastases* may be of almost infinite variety and like those considered in the preceding paragraph may often be the first to appear even though they indicate that the disease is far advanced and beyond more than palliative treatment. Bizarre ways in which this disease can present include pain and swelling of a knee or other joint from secondary deposits abdominal pain simulating a surgical emergency from rapid enlargement of a liver full of metastases and the development of tumours in the skin which may ulcerate The occurrence of secondary deposits in the brain is a common feature of advanced disease but may also be the first evidence of the condition Not infrequently the primary growth in the lung may be difficult to identify and the decision as to whether a brain tumour is a primary one which should be treated surgically or a secondary deposit from a not very obvious bronchial carcinoma is one which often confronts neurologists *Peripheral neuropathies* of various types have special interest in relation *to the early diagnosis of carcinoma of the bronchus and are* considered separately in the section on some special problems *Pulmonary osteo arthropathy* which may be confused with rheumatoid arthritis will also be considered in the next section

PHYSICAL EXAMINATION

As has been pointed out already it is only very rarely that early diagnosis will be based on physical examination of the chest It almost always depends on the recognition that certain symptoms call for further investigation in the first place radiography Nevertheless physical examination of the patient is of the utmost importance It is often possible to decide promptly from the presence of evidence of distant metastases or from other observations that the disease has already passed the early stages and the patient may at any rate be saved the discomfort of the further investigations necessary in preparing for the treatment of localized disease Attention must be directed especially to the presence or absence of superficial lymph gland enlargement the most frequently affected group being that above the clavicle

aspiration pneumonia—a common condition in patients with chronic cough and sputum These infections may respond well to treatment with antibacterial drugs but even when this occurs a chest radiograph should be taken after clinical recovery The special problems of pneumonia in middle aged and elderly patients are discussed again later

It is uncommon except in old people for *general symptoms* such as malaise lassitude and loss of weight to occur early in this disease Indeed the lack of any general disturbance of health in the presence of other evidence suggestive of the condition may be held to support the diagnosis rather than the reverse It is certainly wrong to wait for obvious signs of illness before thinking of diagnosing carcinoma of the lung to do so is to wait until the disease is far advanced The exception to this is that sometimes in old people general deterioration in health with loss of weight and of appetite may precede respiratory symptoms

Symptoms which are produced by *spread of the tumour* from the lung to other structures in the chest are not strictly concerned in the early diagnosis of bronchial carcinoma Nevertheless these symptoms are often those which are first noticed by the patient and it is important to recognize them as early as possible because palliative treatment with X rays may offer some relief provided the disease has not progressed too far Thus early warning of the development of superior vena caval obstruction may be a feeling of fullness in the head especially when the patient bends to tie his shoe laces Usually symptoms of spread to the mediastinum such as hoarseness from involvement of the left recurrent laryngeal nerve dysphagia from obstruction of the oesophagus or the development of the full picture of obstruction of the superior vena cava indicate that the disease is already beyond treatment of any sort In connection with this group of symptoms spread or metastasis to the pleura with the production of pleural effusion should be mentioned briefly The cardinal symptom of this is dyspnoea and although never an early feature of the illness it should be recognized because it is always worth while to treat it Simple aspiration of the fluid will always give relief sometimes for quite long periods of time and when the fluid recurs it may be possible to delay or even prevent its

related to a carcinomatous bronchial block These pneumonias may be treated successfully with antibacterial drugs but the patients should always be radiographed afterwards Even when radiological clearing has occurred if the patient is in good general condition it is usually wise to carry out bronchoscopy because it is certainly possible for the radiograph to return to normal after a pneumonia even when the relevant bronchus is partly occluded by a carcinoma It is especially by careful investigation of this sort of patient that really early diagnosis will be achieved In some patients these pneumonias do not clear up either clinically or radiologically In them the problem of what is called unresolved pneumonia arises It must be accepted that this term is in no sense a diagnosis and although it is true that some uncomplicated pneumonias are slow to clear it is wise to work on the assumption that the cause of a persistent radiographic shadow may be either an empyema a bronchial carcinoma or tuberculosis Further investigation is required in all these patients In middle aged and elderly men the diagnosis of bronchial carcinoma is always an urgent possibility in these *circumstances and bronchoscopy should be carried out as soon* as possible

During mass radiography or other routine radiological investigations it is not uncommon to find in some radiographs rather clear cut shadows in the lung fields Although sometimes the possibility of more uncommon tumours arises in general the main question is whether these *solid lesions* are tuberculous or are peripheral bronchial carcinomas Sometimes it may be possible to settle the question with fair certainty as for example when acid fast bacilli can be demonstrated in the sputum Very often these patients are truly free from symptoms and have no sputum If calcification can be demonstrated in these lesions (and this may sometimes be obvious only on tomography) then carcinoma is not a likely diagnosis Again if these lesions can be shown to be surrounded by one or more neighbouring smaller similar lesions and this may also be obvious only on tomography then a diagnosis of tuberculosis is likely However very often—and this again applies especially to men of middle age and more—unless definite evidence can be produced that such a lesion is tuberculous it is wise to proceed without delay to thoracotomy

and behind the sternal end of the sterno mastoid muscle The presence of hepatic enlargement and the detection of abnormal signs on examination of the central nervous system will often give clear evidence of the presence of metastases In the examination of the chest itself the presence of generalized wheezing may be of more importance in the planning of treatment than the detection of localized signs Only in very advanced disease in very ill patients is it satisfactory to rely on physical signs in the diagnosis of bronchial carcinoma to diagnose early disease radiography must be used and the disease is more likely to be early if localizing signs are not found in the chest

Finger clubbing is frequently associated with bronchial carcinoma and its presence does not necessarily indicate far advanced disease It does occur with other respiratory disease especially suppurative conditions but when it is found it must always direct attention to the chest and point to the need for further investigation The condition called *hypertrophic pulmonary osteo arthropathy* is less commonly seen than clubbing and when found must strongly suggest the presence of a bronchial carcinoma This consists often of pain in the arms or legs usually in relation to the joints In some the joints may be swollen there may be for example effusions in the knee joints Sometimes the hands may be swollen and the fingers stiff and painful This condition may be regarded as an arthritis possibly rheumatoid in type but the observation that the fingers are clubbed—and finger clubbing always accompanies the other manifestations—will suggest the correct diagnosis Pulmonary osteo arthropathy is rarely associated with the relatively early stages of bronchial carcinoma but its recognition does not invariably mean inoperability Moreover removal of the growth or possibly when this cannot be done division of the vagus nerve offers the best chance of relieving this distressing condition

SOME SPECIAL PROBLEMS

There are some points which are worthy of special consideration in patients who are diagnosed as having *pneumonia* In middle aged and elderly men it is a diagnosis that should always cause the doctor to consider whether the consolidation may be

radiography should not be undertaken of the most susceptible group in the population namely heavy cigarette smokers over the age of forty If this were to be successful it would need to be repeated at six monthly or even three monthly intervals and it is doubtful whether this might not in general produce rather than alleviate anxiety However in some individuals regular radiography may be justifiable and some men who have been heavy smokers now ask for routine chest radiographs

METHODS OF INVESTIGATION

The methods which are most useful in the investigation of patients suspected of having bronchial carcinoma are *chest radiography bronchoscopy* and the *examination of the sputum for carcinoma cells* Each of these methods has its limitations It is uncommon but not impossible for the chest radiograph to be completely normal in the presence of a bronchial carcinoma If the tumour is peripheral it may be shown as a shadow of almost any shape and size but is often more or less circular with well defined margins This sort of lesion may break down and present as an abscess which characteristically has a thick and irregular wall Partial obstruction of a major bronchus will produce at first obstructive emphysema and cause the mediastinum to move away from the affected side in full expiration—a feature most obvious when the partial obstruction involves the main bronchus to one lung When complete obstruction occurs then collapse and consolidation of the affected lobe or segment is seen Sometimes tumours in the hilum of the lung can be seen as masses projecting from the mediastinum These appearances may be found with relatively early and removable tumours pleural effusions massive mediastinal glandular involvement and erosion of ribs indicate advanced disease It must also be remembered that it is possible for a small tumour to be present in one of the major bronchi without radiological evidence This is the reason why bronchoscopy should be carried out in certain circumstances even when the radiograph is normal These circumstances have been referred to previously and they are essentially those which strongly suggest the presence of a bronchial carcinoma Thus a patient who has had haemoptysis

A few lesions will be removed which are in fact tuberculous but the danger of waiting until the diagnosis becomes evident is very great and this chance of really early treatment should not be thrown away

There are some types of *neuropathy* which may be the first evidence of the presence of a bronchial carcinoma They are not necessarily associated with advanced growths and when they are recognized the possibility of a bronchial carcinoma must be considered and further investigations begun They form a group of greater interest in relation to early diagnosis than do those neurological complications of bronchial carcinoma which are caused by carcinomatous deposits within the nervous system and which it need hardly be said always indicate that radical treatment is impossible In carcinomatous neuropathy the lesions in the nervous system are not directly caused by deposits of carcinoma Most of the examples reported have been associated with bronchial carcinoma but other growths for example genital and breast carcinomas have also been found Mixed sensory and motor neuropathies are commoner than those which present purely sensory and purely motor disorders The symptoms include ataxia burning and smarting of the limbs with peripheral sensory loss and muscular atrophy and loss of reflexes Sometimes more purely sensory or more purely motor forms have been described Another variety is that known as subacute cortical cerebellar degeneration This type of cerebellar degeneration has several causes it can for example be hereditary but it may also occur in association with carcinoma The symptoms are of fairly rapid progression and do not usually remit They consist of ataxia of gait and later of the hands and speech Nystagmus and less commonly diplopia may be found Vertigo and sensory changes have been described as have pains in the legs and mental deterioration Although these neuropathies are uncommon associates of bronchial carcinoma their symptoms may precede those of the cancer and it is certainly wise to include chest radiography among the investigations considered necessary in patients with neuropathy of obscure origin

It must be accepted that if diagnosis is to depend on symptoms then truly early recognition of bronchial carcinoma will not often be possible The question therefore arises as to whether *routine*

radiography should not be undertaken of the most susceptible group in the population namely heavy cigarette smokers over the age of forty If this were to be successful it would need to be repeated at six monthly or even three monthly intervals and it is doubtful whether this might not in general produce rather than alleviate anxiety However in some individuals regular radiography may be justifiable and some men who have been heavy smokers now ask for routine chest radiographs

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either a definite single episode or repeated streaking of the sputum should certainly be bronchoscoped even if the radiograph is normal Middle aged men who have been heavy smokers and who have had pneumonias of segmental or lobar distribution should be bronchoscoped even though the pneumonia clears up and leaves an apparently normal radiograph

Where possible the clinical and radiological diagnosis of bronchial carcinoma should be confirmed histologically Bronchoscopy is therefore usually undertaken whenever there is clinical or radiological evidence of a growth This may provide further information of value in planning the treatment, and may also allow biopsy to be carried out. Sometimes when the combination of radiological and clinical evidence not only strongly suggests the diagnosis but also indicates clearly that metastasis has occurred and it seems that no treatment is possible it may be reasonable to omit this step on the grounds that even if the diagnosis is proved histologically there is nothing to offer the patient Especially therefore when it seems likely that early diagnosis may be achieved this examination should always be done With the bronchoscope evidence of tumours in the main bronchi and the lobar bronchi can be seen and the orifices of the segmental bronchi can be inspected The method is therefore very limited in its applicability and a positive result is not usually obtained in more than a third of patients with bronchial carcinoma A negative result varies in its value according to the circumstances if the pneumonia under investigation is of lobar extent then a normally patent lobar bronchus suggests that the cause is not a carcinomatous occlusion on the other hand a segmental lesion may be caused by a growth deep inside the segmental bronchus and not visible at the orifice so that negative bronchoscopy may mean nothing Peripheral tumours also are not usually associated with abnormality of the proximal bronchi

When the tumour is beyond the reach of bronchoscopic vision or when histological examination of material removed at bronchoscopy fails to show tumour tissue examination of the sputum for carcinoma cells may establish the diagnosis Where for any reason bronchoscopy cannot be carried out the demonstration of carcinoma cells in the sputum may be the only method of making a final diagnosis There has recently been a good deal

of interest in methods of examining the sputum for malignant cells and pathologists with special experience can achieve positive results in about eighty per cent of patients with bronchial carcinoma False positive findings seem impossible to avoid completely but may be no more than two per cent of the positive results On the other hand a negative finding cannot be regarded as excluding the diagnosis and may mean only that the minute portion of sputum chosen for examination does not contain any cells which can be recognized as malignant A series of examinations therefore gives a more reliably negative result It is however a very time consuming examination and may not be practicable for large numbers of patients Although it is best not to rely on it when material can be obtained for histological examination it may provide in combination with clinical and radiological evidence a means of diagnosing bronchial carcinoma when bronchoscopy fails

Finally it may be said that the best hope of diagnosing bronchial carcinoma reasonably early is to think of it. In all middle aged people men in particular and especially heavy smokers the development of any respiratory abnormality must be regarded seriously and a radiograph should be taken without delay

CHAPTER XI

ENDOCRINE DISORDERS

By D M Dunlop

DIABETES MELLITUS

IN the old days the problem confronting those interested in the management of diabetic patients was how to keep them alive for a while and apart from gangrene we were not much concerned with the complications of the disorder for few patients had lived long enough to develop them Now the problem is not so much that of the acute complications such as diabetic coma for we know or ought to know how to avoid that catastrophe as it is that of a chronic disease with a considerable expectation of life which involves a prolonged struggle to prevent its late complications which are so distressingly common—angiopathy retinopathy nephropathy and neuropathy If we believe that meticulous and aggressive treatment of the metabolic abnormality is at least a factor in preventing or delaying the development of these complications then early diagnosis of the condition is a matter of prime importance It follows that the testing of the urine for glucose should be an invariable part of the routine clinical examination of every patient This sounds a platitudinous piece of advice but it becomes less so when it is remembered that for every known diabetic—and there are about 250 000 in this country—there are probably an equal number of undiagnosed cases The qualitative test for glycosuria—the dipping of a Clinistix paper into a specimen of urine which changes colour in the presence of glucose—is so rapid delicate and specific that there is no excuse for its omission

The classical symptoms of diabetes are of course polyuria thirst, weakness and loss of weight. In the young diabetic such symptoms are nearly always present and may develop acutely so that the time of onset of the disease can be ascertained with

considerable accuracy The discovery of glycosuria in association with such symptoms—and especially when acetone is also present in the urine—makes the diagnosis certain without further investigation It must be remembered however that diabetes only starts under the age of forty in about twenty per cent of cases almost equally divided between males and females In the remaining eighty per cent the disease starts after the age of forty with a considerable preponderance of females owing to the greater incidence of obesity in post menopausal women—and obesity is a potent factor in the aetiology of diabetes In these older diabetics who constitute the great majority of the total the onset of the disorder is often very insidious and symptoms may be minimal for many years In consequence what brings such patients to the doctor may be the development of one of the complications of the disease rather than polyuria thirst weakness and loss of weight Thus pruritus vulvae boils or carbuncles neuritic pains in the legs lesions on the toes or feet due to a mixture of diabetic angiopathy and neuropathy massive gangrene failing vision as the result of retinopathy or cataract oedema due to diabetic nephropathy (the Kimmelstiel Wilson syndrome) or the development of tuberculosis to which the diabetic is so prone may be the first presenting sign or symptom in the older patient

When symptomless glycosuria is discovered a blood glucose determination should be made on a specimen procured two hours after a normal breakfast If the value is below 120 mg per 100 ml the patient is almost certainly not a diabetic If it exceeds this figure a full glucose tolerance test should be carried out which will help to distinguish diabetes mellitus from the many other causes of glycosuria In the normal person the fasting blood glucose concentration seldom exceeds 100 mg per 100 ml and after 50 g of glucose have been ingested it returns to its fasting level in two hours time It is important to remember that the previous diet may influence the form of the glucose tolerance curve since the state of the subject s carbohydrate nutrition affects the response to exogenously administered glucose An adequate store of glycogen in the liver is essential for a normal response to be obtained Thus after a low carbohydrate high fat diet—or still more after complete starvation—the rise in

the blood glucose concentration following the ingestion of glucose is more pronounced and the fall delayed It is not uncommon for a person in whom glycosuria has been discovered at a Life Assurance Examination to restrict drastically his carbohydrate intake if he is then subjected to a glucose tolerance test the result may suggest that he is a mild diabetic even though his initial glycosuria may have been due to other causes It is therefore most important to ensure that an ordinary mixed diet has been taken for some days prior to the test

There are of course many explanations for hyperglycaemia or glycosuria besides diabetes and when glycosuria is discovered it is essential to keep the other possible causes in mind before diagnosing diabetes mellitus The true cause may often be obvious enough from the history and clinical examination in others a glucose tolerance test may be necessary to establish the diagnosis

Sporadic or continuous glycosuria occurs in about twenty five per cent of cases of hyperthyroidism and since weakness loss of weight and some thirst may also be present it is not unusual for hyperthyroid patients without prominent goitres or exophthalmos to be wrongly diagnosed as suffering from diabetes

Adrenergic influences are responsible for the hyperglycaemia which may accompany states of excitement anger or fear and the clinical analogues of Claude Bernard s classical experimental puncture of the floor of the fourth ventricle in rabbits find their expression in the glycosurias which not infrequently occur following fracture of the skull tumours of the base of the brain intracranial haemorrhage meningitis epilepsy and encephalitis

The hyperglycaemic effects of anterior pituitary and adrenocortical hormones are well known and it is not therefore surprising that hyperglycaemia occurs in about fifteen per cent of cases of acromegaly and in a large percentage of patients with Cushing s syndrome Further the administration of cortisone or corticotrophin may not only exacerbate an existing diabetes but may induce a temporary hyperglycaemia and glycosuria in patients not previously diabetic It is probable that when this occurs it is indicative of a pre-clinical diabetic state

A decreased glucose tolerance is found in about seventy five per cent of cases of haemochromatosis (bronzed diabetes) due to

degenerative changes induced by the deposition of haemosiderin in the pancreas The same effect is produced in a few cases of pancreatitis and in about thirty per cent of patients suffering from carcinoma of the pancreas—particularly when it affects its body and tail

If the absorption of glucose from the intestine is unduly rapid the blood glucose concentration may rise to an unduly high level before glycogenesis has time to overtake it and hyperglycaemia and glycosuria may occur but the very height of the glucose concentration then stimulates rapid glycogenesis and the quick rise of the blood glucose concentration to a high peak is followed by a precipitous fall Such hyperglycaemia followed sometimes by hypoglycaemia is not uncommon following subtotal gastrec tomy in which food reaches the jejunum with undue rapidity

Lastly a lowering of the renal threshold for glucose which is very apt to occur in pregnancy may result in a harmless functional glycosuria readily distinguishable from diabetes by the normal glucose tolerance curve The condition of low threshold glycosuria is less common than was once supposed and there is real danger of confusing an early mild case of diabetes with it unless the glucose tolerance test gives absolutely unequivocal evidence in favour of renal glycosuria Even then it is wise to repeat the test after an interval before the diagnosis of low threshold glycosuria is established On the other hand patients with this harmless abnormality have often been stigma tized as suffering from diabetes with all the inconvenience that such a diagnosis implies including difficulty in securing life assurance

MYXOEDEMA

Patients suffering from myxoedema often have to wait for a long time before their malady is correctly diagnosed Primary atrophy of the thyroid as distinct from hypothyroidism result ing from thyroidectomy antithyroid drugs or pituitary in sufficiency occurs very slowly and insidiously and is usually encountered in elderly people The practitioner who may see such patients at frequent intervals gets used to the gradual altera tion in their physical and mental state and is apt to ascribe any changes which he does note to the natural development of old

age and senility Few doctors can have escaped the humiliating experience of having the true diagnosis pointed out by some colleague seeing the patient for the first time to whom the clinical features of myxoedema may be immediately obvious

It is most important to make the diagnosis at an early stage of the disorder for under such circumstances myxoedema is not only one of the most satisfactory and easy diseases to treat—it is almost as though the wand of a fairy has passed over mind and body when thyroid extract or thyroxine are given in appropriate amounts—but in addition long continued untreated myxoedema has as its almost invariable concomitants grave disturbances of the heart and coronary vessels the myocardium becomes soft flabby and dilated and the coronary arteries atherosclerotic and in such cases any attempt to give adequate replacement therapy may result in cardiac decompensation the practitioner may thus have to content himself by alleviating only the grosser manifestations of hypothyroidism steering a difficult course between the Scylla of myxoedema on the one hand and the Charybdis of effort angina on the other

All patients suffering from myxoedema feel weak and lethargic, but these are such non specific and common symptoms that they are of little value in differential diagnosis The muscles become stiff and the movements slow and awkward

The skin is almost invariably dry cold and rough Its texture resembles the hands of women which are frequently in the wash tub It tends to become infiltrated with the typical myxoedema tous fluid which does not pit on pressure and which is usually particularly obvious in the peri orbital region The sweat and sebaceous glands are diminished and as many myxoedematous patients become careless in their habits accumulations of desquamated skin and dirt may form thick bark like crusts especially on the feet The nails are short atrophic and brittle Baldness is common and the body hair sparse but it must not be thought that all patients lose their scalp hair as this only becomes scanty in about sixty per cent of cases it is almost invariably coarse and dry There is decreased sweating and an undue sensi tivity to cold so that the patients wear masses of clothes and scorch their legs by sitting as close as possible to the fire in this country therefore it is exceptional to find a myxoedematous

woman without the marks of erythema ignis on her legs The thickness of the skin the diminished amount of blood flowing *through it and the mild anaemia which characterizes the condition* all go to produce a pale skin sometimes tinged with yellow due to carotinaemia the result of the very slow metabolism of carotene to vitamin A The face tends to be expressionless apart from a vaguely puzzled look

The thick tongue the infiltration of the larynx with myxomatous material and the weakness of its intrinsic muscles account for the highly characteristic thick croaky voice of the patient

Though exertional dyspnoea is common ordinary clinical examination of the cardiovascular system in myxoedema does not often reveal any characteristic diagnostic features though on radiological examination the heart in advanced cases assumes the shape of an association football owing to dilatation of all its chambers and to the presence of some fluid in the pericardium in addition the electrocardiogram shows a decrease in the amplitude of its complexes

Intractable constipation is present in about sixty per cent of cases and the abdomen is usually distended In some fifty per cent of patients the appetite becomes very poor and some cannot bestir themselves to cook proper meals this accounts for the fact that myxoedematous patients—contrary to common belief—are by no means always fat in spite of their slow metabolism

Slowness in cerebration and in emotional reactions are almost invariable features of myxoedema and a poor memory is common On the whole the patients tend to be rather good natured people with a slow dry humour The late stages of the disease may be associated with myxoedematous madness and ultimately coma

The diagnosis of well developed myxoedema once the disease is thought of presents little difficulty and laboratory diagnostic aids are unnecessary In the earlier stages of thyroid insufficiency however it may be difficult to differentiate it from senile changes and from certain psychoses Myxoedema is also occasionally confused with nephrotic nephritis and with pernicious anaemia

In *myxoedema and cretinism the blood cholesterol concentration* is practically never below 200 mg and may even exceed 500 mg per 100 ml Thus in a patient exhibiting a number of

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hardly detectable by the most expert examination that there may be no eye signs at all—or alternatively that exophthalmos may occur without thyrotoxicosis and that in some cases only a few of the classical signs and symptoms may be obvious Medical students often say this patient can t be thyrotoxic because she hasn t lost weight or because she hasn t got a fast pulse or because she says she is not nervous We must realize that the textbook picture of most diseases is a composite one made from the study of many patients but that the individuals who make up the composite picture may differ from it widely in many respects We have no more reason to expect to be presented in disease with a royal straight flush of signs and symptoms in every case any more than we can expect to hold a royal straight flush in every hand at poker Thus a girl may have a small goitre without exophthalmos and may give a vague history of ill health Is her goitre simple or is it mildly toxic? A young soldier complains of sweating tachycardia palpitation and nervousness without obvious thyroid enlargement or eye signs Is he suffering from the effort syndrome or is the true cause of his symptoms hyperthyroidism? An elderly woman has unexplained auricular fibrillation with cardiac decompensation and a somewhat raised blood pressure she has practically no other signs or symptoms of thyrotoxicosis apart from the doubtful presence of a small nodule in her thyroid Is she suffering from thyrotoxic or hyper tensive heart disease? These are diagnostic problems of great difficulty which must confront every practitioner not infrequently

Early diagnosis is of great importance for the treatment of thyrotoxicosis is exceedingly satisfactory and most patients can be restored to good health but if it is allowed to progress un treated for a long time permanent cardiovascular and often psychological damage are bound to ensue Further if undiag nosed untreated thyrotoxic patients are subjected to stress—such for instance as a surgical operation or a severe pneumonia—they may be precipitated into dangerous and sometimes fatal thyro toxic crises In a book of this kind it is undesirable to attempt to detail all the protean manifestations of thyrotoxicosis which may be encountered but in the author's view the following are the most important points to emphasize in the clinical diagnosis of the condition

the signs and symptoms which have been mentioned a high blood cholesterol level is practically diagnostic of the condition provided Type II nephritis can be excluded which should not be difficult It is a particularly valuable test for cretinism or juvenile myxoedema as in babies and children determinations of the B M R are impracticable and radioactive iodine tests contraindicated

The basal metabolic rate is reduced in myxoedematous patients to between minus twenty and minus forty per cent of normal Unless carefully performed the test has numerous fallacies and may be most misleading but a result below minus fifteen is of much greater diagnostic significance than one suggesting a slightly increased metabolic rate

The diagnosis of myxoedema can be confirmed with a high degree of accuracy by studies with tracer doses of radioactive iodine (see pp 161 2) Facilities for such specialized procedures are not as yet universally available and a therapeutic diagnostic test with thyroid extract or thyroxine may then be justifiable if the patient has myxoedema a clear cut clinical response should be obvious in a fortnight

HYPERTHYROIDISM

No attempt will be made in this section to classify thyrotoxicosis into different pathological types Possibly any attempt to do so is highly artificial as one type merges almost imperceptibly into the other and the difference in the clinical features which occur may simply represent the effect of age in modifying the condition

Thirty or forty years ago most people thought the diagnosis of thyrotoxicosis was very easy and its treatment very difficult now most of us realize that in some cases the diagnosis may be most complicated though the treatment is much more satisfactory The diagnosis is of course perfectly simple when a patient has exophthalmos thyroid enlargement tremor sweating hypersensitivity to heat tachycardia fatigue and weight loss it is probable that in the old days the condition was seldom diagnosed unless most of these features were present We now know that thyrotoxicosis may be present with minimal thyroid enlargement

mias are common The tachycardia when present tends to persist during sleep in contrast to the slower sleeping pulse of patients who suffer from functional tachycardia When there is no obvious explanation for tachycardia or for a cardiac arrhythmia hyper thyroidism must always be considered as a possible cause

Thyrotoxicosis is one of the common reasons for unexplained loss of weight though this is not an invariable feature of the disorder *Further the increased metabolism may stimulate the* appetite so that the likelihood of hyperthyroidism being present is greatly increased when weight loss is associated with a good appetite Diabetes is the only other condition in which this association occurs at all commonly

Only a small percentage of hyperthyroid patients suffer from actual diarrhoea but on the other hand they are practically never constipated Do you take frequent laxatives? should be another invariable diagnostic question and an affirmative answer is a strong point against a diagnosis of thyrotoxicosis

In spite of the mass of clinical evidence which in any given case can be marshalled in favour of or against a diagnosis of thyrotoxicosis it must be confessed that in a few cases the evidence is so equally balanced on both sides as to leave the diagnosis in doubt and recourse must be made to laboratory aids Persistently high values of the basal metabolic rate—above twenty per cent of normal—when skilfully carried out are diagnostically significant though it is just in the doubtful borderline case where such help would be particularly useful that the results of the test may be equivocal a single determination on a nervous out patient may be most misleading and is worse than useless The determination of the protein bound iodine of the plasma furnishes a specific means of measuring the amount of circulating thyroid hormone and so of assessing the activity of the gland Unfortunately the determination of such minute quantities of iodine is a delicate and difficult chemical procedure which is seldom available for routine use Fortunately it is possible to obtain related information more easily and to trace in some detail the fate and distribution of iodine in the body by the use of the radioactive isotope of iodine ^{131}I and when facilities are available for such studies a diagnosis can be made with a high degree of precision In the absence of previous recent

Almost all thyrotoxic patients are nervous Nevertheless nervousness is so common and is indeed an attribute which patients suffering from almost any condition seem anxious to arrogate to themselves that the symptom may not by itself be of much value in differential diagnosis The nervous signs and symptoms of thyrotoxicosis however have certain characteristic features If the patient is watched her movements tend to be quick and jerky It is instructive to ask her to button her blouse or pyjama jacket when her hyperkinetic rapid clumsy movements are often reminiscent of patients suffering from chorea The tremor of the extended hands which is usually present, is fine and rhythmical—unlike the coarser tremor of other nervous people The thyrotoxic patient is usually a difficult person to live with she constantly loses her temper with her children and husband and then overcome with remorse bursts into floods of tears Weeping is usually a common feature of the condition and the patient is often at a loss to know what she is weeping about Every molehill becomes a mountain to the thyrotoxic whose neurosis often takes the form of an excessively anxious solicitude for her family in spite of her exhaustion she feels impelled to busy herself continually in an ever changing series of activities which are often pathetically ineffectual

The vast majority of hyperthyroid patients are hypersensitive to heat and sweat excessively The skin of such patients is moist warm fine and flushed the hand has the texture of a child and the feel of it is of the greatest diagnostic importance It is far more instructive to hold hands with thyrotoxic patients than to determine their basal metabolic rates Though occasionally their hands may be cold and wet like frogs this is much more a feature of the effort syndrome than of thyrotoxicosis If a patient is found to be wearing a heavy coat a thick jersey or a woolly bedjacket even on a cold day it is a point against the diagnosis Do you feel the cold much? is a diagnostic question which should never be omitted The thyrotoxic will usually answer with an emphatic negative

Exertional dyspnoea and palpitation are common symptoms in association with a hyperkinetic circulation The pulse tends to be rapid with a raised pulse pressure the apex beat is forcible and in older patients auricular fibrillation or paroxysmal arrhyth

patients are notoriously intolerant) a mild infection or a minor surgical operation

Incidence of Common Symptoms and Signs in Addison s Disease

	per cent
Tiredness and weakness	100
Loss of weight	100
Skin pigmentation	97
Blood pressure below 110/75	90
Gastro intestinal symptoms	80
Early morning hypoglycaemia	80
Buccal pigmentation	73
Attacks of reactive hypoglycaemia	30

The commonest presenting symptom in Addison s disease is a feeling of weakness and tiredness Darkening of the skin may be the first sign to attract attention especially in women though men of the labouring class may fail to notice a degree of pigmentation which may be obvious to an outside observer Sometimes such patients come to the doctor because of anorexia and vomiting and occasionally because of extreme mental depression

Whatever the presenting symptom a feeling of tiredness and weakness is an invariable feature The only other absolutely constant finding is loss of weight sometimes amounting to only a few pounds but usually to a considerable amount Anorexia nausea and occasional vomiting are common Most patients are hypoglycaemic in the early morning being difficult to rouse from sleep and pathologically surly and resentful towards those who attempt to awaken them A few react violently to the unopposed endogenous insulin which they secrete in response to a carbohydrate meal so that in about an hour and a half after food they become sleepy weak and faint

Extreme hypotension is not an essential feature of the disease but a systolic pressure above 110 in an untreated patient is a strong point against the diagnosis and one above 120 practically excludes it No other single physical sign is as valuable as pigmentation in diagnosis for some degree of it is almost invariable It varies from the faintest olive tinge somewhat reminiscent

treatment with iodides or other antithyroid drugs the rate of uptake of ^{131}I by the gland is an index of thyroid function Thus in hyperthyroidism an overactive gland takes up—as measured by an appropriate counter—from forty to ninety per cent of the tracer dose in a few hours whereas in myxoedema less than fifteen per cent is very slowly absorbed by the gland In the normal person the curve of uptake lies between these two extremes Conversely the excretion in the urine of thirty per cent or less of the tracer dose of ^{131}I in forty eight hours indicates increased avidity of the gland for iodine or hyperthyroidism whereas a high excretion of from seventy to ninety per cent indicates hypothyroidism Again the normal person gives inter mediate results Lastly if the protein bound plasma radioactivity forty eight hours after the tracer dose exceeds 0 4 per cent of the dose per litre of plasma thyrotoxicosis is almost certainly present When the apparatus is available this latter test is not only an accurate but a very simple one to have carried out in practice as it only involves the patient in drinking the tracer dose and then forty eight hours later the doctor removes a sample of blood which is sent to the laboratory

ADDISON'S DISEASE

Addison s disease is a state of chronic adrenal cortical in sufficiency resulting from tuberculous destruction or idiopathic atrophy of the cortex of both adrenal glands It is not synonym ous with cortical insufficiency which may follow failure of the anterior pituitary to secrete its adrenocorticotrophic hormone Cases are fairly evenly distributed between males and females The disorder seldom occurs before the age of fifteen or after fifty five In the old days early diagnosis of the condition did not matter in fact the later the diagnosis was made the better as it implied a death warrant for the patient Nowa days early diagnosis is a matter of crucial importance for with efficient modern replacement therapy patients can be restored to an active healthy and vigorous existence whereas untreated patients are sure to die within two years often being precipitated into fatal Addisonian crisis by some form of stress which may be quite trivial—an injection of morphine (to which drug such

factors one of which is an adequate supply of adrenal gluco corticoids In Addison s disease this copious diuresis does not occur and the response to fluid administration is paradoxical the volume of urine secreted during the night exceeding that passed during the day

A more direct assessment of the functional activity of the cortex is obtained by estimating the urinary excretion of neutral 17 ketosteroids which reflects the level of secretion of sex hormones by the adrenal cortex and to a less extent the testis The excretion of less than four mg of these substances in twenty four hours is almost invariable in cases of Addison s disease but the test is only specific in the negative sense since the excretion of larger amounts makes the diagnosis untenable

In people with normal adrenal cortical function a profound fall in the level of circulating eosinophils follows the intra muscular injection of corticotrophin being most marked four hours after the injection The phenomenon depends on an increased endogenous production of cortical hormones If no significant fall in the eosinophil count occurs it is reasonable to infer that no functional adrenal cortical tissue exists to respond to the stimulus

All patients with Addison s disease are profoundly sensitive to insulin and the intravenous injection of not more than two units of soluble insulin results in them in a precipitous fall in the blood glucose to hypoglycaemic levels which is not recovered from in two hours time Facilities must always be at hand to cut short an excessively severe and possibly dangerous hypo glycaemic reaction to such a test

Addison s disease is most frequently confused with asthenic neurasthenia carcinoma of the stomach and other conditions giving rise to a somewhat similar pigmentation such as haemo chromatosis and vagabond s disease

ANTERIOR PITUITARY INSUFFICIENCY

A greater or less degree of anterior pituitary insufficiency is a more common cause of chronic individualism than most people suppose In the past it was seldom recognized except in its most florid forms We have become more conversant with its clinical

of the sun kissed skin of the Mediterranean peoples to a dark brown almost negroid hue The pigmentation is most conspicu ous on the exposed parts, on areas subjected to pressure on the nipples and in the flexures It is much more marked on the back of the hand especially over the knuckles and this dark colour contrasts—as in Indian races—with the comparatively pale palms where the pigment is confined to the skin creases. Occasionally the pigmentation alternates with leucodermic pat terns Patches of pigmentation on the mucous membrane of the *inner surface of the lip and cheek gums hard palate* and tongue if not pathognomonic of Addison s disease are nearly so Such buccal pigmentation however only occurs in patients in whom the skin is deeply pigmented and not in less typical cases where its presence would be most helpful in diagnosis

Some seventy per cent of patients suffering from Addison s disease conform to the classical pattern which has been described In such cases once the possibility has been entertained of this rare disease being present the diagnosis presents no difficulty and it is amply confirmed by the dramatic response of the patient to appropriate treatment The diagnosis of Addison s disease must be less easy in parts of the world where the physiological causes of skin pigmentation are more common than in this rather sunless country and it must be very difficult in the case of an asthenic dyspeptic hypotensive Indian

A variety of diagnostic tests are available for doubtful cases In a patient with suggestive symptoms and signs the radiological demonstration of calcification of the adrenal glands gives un equivocal proof of the diagnosis of Addison s disease and of its tuberculous aetiology though of course a negative X ray does not exclude it There are in addition tests which indicate deficiency of the mineralo corticoids and yet others which give a rough assessment of deficiency of the cortical sex hormones

Estimation of the serum level of sodium and chloride is a simple procedure and though normal figures are quite compat ible with untreated Addison s disease low values are at least suggestive They acquire greater significance if they can be shown to be associated with a normal or high excretion of sodium and chloride in the urine The prompt diuresis which normally follows the giving of a large quantity of fluid depends on a number of

The tests already described for the diagnosis of hypothyroid ism and adrenal cortical failure may all be found to give positive results in pituitary insufficiency In addition the secretion of the anterior lobe hormones is absent or defective The assay of these in the blood or urine is still mostly in the experimental stage the one which can be estimated with least difficulty is the follicle stimulating hormone little if any is found in the urine in pituitary failure

It is important to distinguish the condition from *primary myxoedema* since pituitary insufficiency demands treatment with cortisone as well as with thyroid indeed if thyroid alone is given to such patients it may make them considerably worse It is usually possible to make the differential diagnosis from the history and from the texture and appearance of the skin which as a rule differs considerably in the two conditions Where doubt exists and facilities for radioactive iodine studies are available an injection of thyrotrophin will cause the uptake of the isotope by the thyroid to be considerably increased in hypothyroidism due to pituitary insufficiency but will have no such effect in primary myxoedema

The other condition which may be confused with pituitary failure is *anorexia nervosa* and indeed long continued sub nutrition has a profoundly depressing effect upon the function of the hypophysis Thus anorexia nervosa is also characterized by amenorrhoea a low rate of metabolism and an undue sensitivity to cold The points of importance in differential diagnosis are as follows

1 The patient with anorexia is commonly young unmarried and has had obvious psychological difficulties in hypopituitarism she is usually older has had children and may have been psychologically stable before the onset of symptoms

2 In anorexia the patient is usually cachectic which is un common in hypopituitarism

3 The patient with anorexia nervosa has often a bird like activity protesting that she is fit for anything and that there is nothing the matter with her profound asthenia is an almost invariable concomitant of pituitary insufficiency

4 The skin in anorexia is blue cold and rough in contrast to its parchment like pallor in hypopituitarism

features since hypophysectomy has been more widely practised for a variety of pathological conditions of the gland and particularly for the treatment of certain malignant metastases It is of great importance to diagnose hypopituitarism at an early stage for once more as in the case of the conditions already discussed modern treatment may not only be life saving but may transform a chronic invalid into a reasonably healthy person

Apart from ablation of the gland for therapeutic purposes pituitary insufficiency may occasionally result from basal meningitis or encephalitis trauma chromophobe adenomata or craniopharyngiomata or it may occur following the successful treatment of acromegaly when hyperpituitarism not infrequently gives place to hypopituitarism Far the commonest preceding history however is that of a severe post partum haemorrhage in which necrosis of the pituitary may result from circulatory obstetric shock (Sheehan s disease) In a shorter or longer time after such an event the clinical features of gonadal thyroid and adrenal cortical insufficiency may arise usually in that order owing to failure of the stimulating effect of the pituitary trophic hormones on the subsidiary glands

Amenorrhoea is a constant feature of the condition associated with atrophy of the genitalia loss of pubic axillary and body hair and failure of libido The signs and symptoms of thyroid and adrenal cortical insufficiency which have already been discussed may be present to a greater or less extent with the exception of course of Addisonian pigmentation since this is produced in Addison s disease by the pituitary melanophore hormone secreted along with corticotrophin in a vain effort to flog into life again the dead horse of the adrenal cortex Far from being pigmented the skin in hypopituitarism is remarkable for its pallor It is seldom myxoedematous in appearance but is more usually thin shrivelled and inelastic and occasionally very soft smooth and shiny A few patients may be cachectic and a few are overweight but great abnormalities in weight are not common features of the disorder Diagnosis usually rests upon the onset of amenorrhoea following a severe post partum haemorrhage in association with loss of body hair and on the characteristic appearance of the patient

which the patients complain is a soft tissue and cartilaginous enlargement of the joints

All the soft tissues viscera and organs become enlarged The skin for instance is two or three times its normal thickness being hypertrophied and coarse and appearing too big for the body This may result in deep corrugations developing over the fore head and scalp as occurs in bloodhounds or bulldogs The hair follicles hypertrophy as do those of the sweat glands which may easily be seen with the naked eye The tongue nose and lips become enormous and the larynx increases in size so that the voice is deep and thick

During the early active phases of acromegaly there is often evidence of increased activity of the thyroid adrenal cortical and gonadal functions Thus there may be increased sweating goitre formation hirsutism a pronounced libido lactation polyuria thirst and glycosuria

The period of active acromegaly may be relatively short and many acromegalics whom we see are burnt out cases in which a state of hypopituitarism has supervened in them all the changes described—apart from the persisting skeletal ones—are reversed and the patient becomes weak and depressed

The fully developed picture of acromegaly is so striking that it is the easiest of disorders to diagnose In its early stages the chief difficulty lies in differentiating patients with the condition from heavily built people with a family appearance of prog nathism who happen to be suffering from a headache due to some other cause Under such circumstances radiological examination of the skull and hands seldom leaves the diagnosis in doubt In addition the B M R is usually moderately raised the responses to a tracer dose of radioactive iodine are those which we associate with mild hyperthyroidism and some twenty per cent of acro megalics show a diminished glucose tolerance occasionally to the extent of having severe diabetes

HYPERPARATHYROIDISM

Primary hyperparathyroidism (generalized osteitis fibrosa or Von Recklinghausen s disease of bone) is due in the great majority of cases to a parathyroid adenoma It is a relatively

5 In anorexia nervosa the axillary and pubic hair is seldom lost to the extent seen in hypopituitarism and there is indeed a tendency for a growth of lanugo like hair to occur on the back

6 The genitalia and breasts do not usually atrophy in anorexia as they do in pituitary insufficiency

ACROMEGALY

Acromegaly which is due to an eosinophilic tumour of the pituitary occurs most commonly about the third decade and has an equal incidence among men and women Its early diagnosis and treatment may prevent not only severe symptoms such as headache but also irreversible and most distressing physical changes including failure of vision

Since the pituitary is surrounded by bone the growth of the tumour is often accompanied by intractable headache due to expansion of the sella turcica In about sixty per cent of cases the tumour impinges on the crossed fibres of the optic chiasma causing bitemporal hemianopia the visual acuity gradually diminishes and total blindness may ultimately ensue but papill oedema seldom occurs

During the active phase of acromegaly there is usually evidence of hyperactivity of all the glands under the influence of the trophic hormones of the pituitary but the most obvious clinical manifestations are of course those produced by excessive secretion of growth hormone Since the disease generally starts after the epiphyses have closed there is no further growth in height and the bones hypertrophy transversely—the thickening being most marked where the epiphyses have fused This is typically seen in the splaying out of the terminal phalanges of the hands and feet The face is broad with a characteristic prognathous appearance the supra orbital region being pro minent due to expansion of the frontal sinuses The lower jaw is enlarged with the teeth widely separated and dorsal kyphosis and lumbar lordosis are commonly encountered often associated with osteoporosis These bony changes result in characteristic radiological appearances—particularly enlargement of the sella and erosion of the clinoid processes The typical acromegalic arthropathy which accounts for many of the aches and pains of

duodenal ulcer The diagnosis cannot be established by clinical examination since it is most exceptional to be able to palpate a parathyroid adenoma and it must depend on biochemical and radiological evidence

The differential diagnosis must include the other causes of nephrolithiasis and renal calcinosis and when symptoms refer able to bone involvement are present osteomalacia osteoporosis hypervitaminosis D multiple myeloma primary and metastatic neoplasms of bone Paget s disease and focal osteitis fibrosa (Albright s syndrome)

The findings of persistently high levels of the serum calcium (between 11 and 20 mg per 100 ml) and of a low serum inorganic phosphate (below 3 mg per 100 ml) are sufficient to establish the diagnosis especially when associated with a raised serum alkaline phosphatase (above the normal range of five to fifteen units) the latter however is only increased when the disease has caused significant changes in the bones When the biochemical findings are equivocal the increased amount of calcium excreted in the urine may be of diagnostic help as this continues to be high even when the patient is given a low calcium diet for some time Calcium balance tests are complicated and time consuming and can rarely be carried out accurately outside the metabolic ward of a well equipped hospital When skeletal changes have occurred the radiographic appearances may show the highly characteristic rarefaction of the whole skeleton in association with multiple cystic areas particularly in the long bones and the moth eaten appearance of the skull

rare disease but since it is eminently treatable it is important to diagnose it early and to attempt to remove the adenoma before serious damage has been done to the kidneys and irreversible skeletal deformities have occurred As the result of the increased secretion of parathyroid hormone calcium is mobilised from the bones into the blood so that excessive amounts of calcium and phosphate are passed in the urine and hypercalcaemia and decalcification of the bones occur Symptoms may be related to the renal tract, to the systemic effects of the hypercalcaemia or to the changes produced in the bones and they tend to occur in that order

As the result of the large quantities of calcium and phosphate passed in the urine polyuria and thirst may occur which when no glycosuria is discovered sometimes lead to a diagnosis of diabetes insipidus If the condition is [illegible] to persist, renal stones of calcium [illegible] or calcium may be deposited [illegible] kidney causing the serious [illegible] result the patient may suffer [illegible] and may eventually [illegible] It is now recognized [illegible] common aetiological [illegible] to be [illegible] may [illegible] The cause [illegible] T[illegible] not [illegible] may [illegible] and [illegible] the [illegible]

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Since hypercalcaemia reduces muscle tone the patient may complain of weakness apathy anorexia and constipation The cause of the frequent association of duodenal ulcer is obscure

The mobilization of calcium from the bones into the blood not only causes decalcification but the normal bone structure may be replaced by cysts composed of fibrous tissue osteoblasts and osteoclasts Consequently the patient experiences pain in the bones particularly in the back and legs and may develop pathological fractures or skeletal deformities In hyperparathyroidism the jaw is sometimes involved by a tumour which closely resembles an epulis.

The symptomatology of hyperparathyroidism is non specific The possibility of its presence should be kept in mind as a cause of nephrolithiasis particularly when this is bilateral or recurrent and when it is associated with weakness bone pain epulis and

metastases will be overlooked Again the pituitary adenoma is now usually diagnosed at an early stage subjected to operation or irradiation and unlikely to reach the post mortem room It is indeed rarely seen by the neurologist

As in other medical fields the pattern changes with the passage of time Although the incidence of cerebral metastases from bronchial carcinoma is steadily increasing in Great Britain the cerebral tuberculoma is disappearing twenty five years ago the tuberculoma constituted a third of all solid intracerebral masses but now the incidence has fallen to one per cent Even so tuberculoma rarely presents as a space occupying lesion and is usually an unsuspected post mortem finding in a patient with tuberculous meningitis Intracranial gumma was always a great rarity even in a highly syphilized population and it must now be regarded as non existent in this country

A recent survey of a consecutive group of space occupying lesions from three different sources included cases from the neurological neurosurgical and neuropathological departments of a teaching hospital If brain abscess and subdural haematoma are excluded the incidence of other lesions shows little variation from one department to another both these conditions are frequently referred direct to the neurosurgeon and the results of prompt treatment in subdural haematoma are so good that such cases rarely reach the neuro pathologist Excluding these two cystic conditions each of which constitutes about ten per cent of the space occupying lesions seen by the neurosurgeon the incidence of glioma is about sixty per cent so that this is the approximate incidence in all those diagnosed as having a cerebral tumour With a similar proviso the incidence of metastases is about one in ten The frequency of meningioma is higher in the neuro surgical series than in the autopsy figures since this is a tumour often amenable to complete surgical removal it again constitutes about ten per cent of clinically diagnosed tumours Apart from the foregoing many other neoplastic and developmental lesions occur but are seen only rarely

The frequency of different forms of tumour varies con siderably with age though most have their favourite sites For example the gliomata are found from infancy to the senium and may involve any part of the cerebrum or cerebellum but one

CHAPTER XII

CEREBRAL TUMOUR

BY HUGH GARLAND

IT is not generally appreciated that tumours are found in the brain as frequently as in most other organs including such sites as the breast stomach lung or rectum Moreover the brain probably shows a wider range of neoplastic changes than any other viscus The clinical picture of a space occupying lesion within the skull may however be produced by other lesions including brain abscess and subdural haematoma and occasionally by processes such as meningitis or thrombosis of intracranial veins and sinuses Even the effects of focal cerebral ischaemia resulting from atherosclerosis may present in this form so that on occasion clinical differentiation may be difficult or even impossible But the clinical objective today is to make a diagnosis which is accurate both as to site and to pathology

Precise figures relating to the incidence of intracranial space occupying lesions are not available and the relative frequency of the different types varies according to the source of the material The reasons for these anomalies are complex but important and include the following It is nowadays the aim of the neurologist and neurosurgeon to prove the existence and the pathology of a tumour beyond all doubt either by exposure and removal or at least by biopsy If the tumour is found to be a glioma of the more rapidly growing kind (glioblastoma) no further action is usually taken and death often occurs at home If the patient appears to be suffering from multiple intracranial masses and has a proved bronchial or other carcinoma he will likewise often die at home without post mortem confirmation Moreover the brain is not always examined at autopsy when a patient has died of malignant disease without cerebral symptoms so that in a post mortem series some

metastases will be overlooked Again the pituitary adenoma is now usually diagnosed at an early stage subjected to operation or irradiation and unlikely to reach the post mortem room It is indeed rarely seen by the neurologist

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The frequency of different forms of tumour varies con siderably with age though most have their favourite sites For example the gliomata are found from infancy to the senium and may involve any part of the cerebrum or cerebellum but one

form (the medulloblastoma) is largely limited to children the frequency of supra tentorial gliomata increases up to middle age after which they become less frequent

The symptomatology of intracranial space occupying lesions covers an enormous clinical field It depends on the age of the patient the duration and nature of the pathological process and its site An adenoma of the pituitary body in an adult results in a syndrome that shares no common ground with a cerebellar glioma in a child or a temporal lobe abscess at any age In approaching the problems of early diagnosis it is therefore necessary to keep in mind a wide variety of clinical manifestations at all ages and including acute subacute and very chronic symptoms or syndromes

In general intracranial tumours present in one of four ways namely with evidence of *increased intracranial pressure epilepsy focal signs and symptoms* and *mental changes* Any one of these may be present alone for long periods or any two or three or all four may appear together These four modes of onset will now be discussed but there will still remain some pathological processes calling for separate description.

INCREASED INTRACRANIAL PRESSURE

Some tumours produce neither signs nor symptoms until they have achieved considerable size but ultimately a tumour will result in symptoms by virtue of its volume alone Since the adult skull can be regarded as a rigid box an addition to its contents can only occur at the expense of other occupants at first the cerebrospinal fluid and later the venous blood It is generally accepted that as intracranial pressure rises certain signs and symptoms result though the mechanism of their production is by no means fully understood The syndrome resulting from raised intracranial pressure (irrespective of cause) is classically regarded as headache vomiting and papilloedema but it is not always as simple as this and each aspect must be considered individually

Of these three manifestations *headache* is certainly the commonest, and in the author's experience it is second only to epilepsy as a presenting symptom of intracranial tumour occurr

ing initially in about a quarter of all cases though ultimately two thirds develop the symptom The mechanism of production of headache is not always clear It is not invariably produced by increased intracranial pressure alone and very high pressures have been recorded without headache Probably more important than the pressure itself is distortion particularly of the dural folds (the falx and the tentorium) and perhaps of the cerebral arteries Headache is always an important diagnostic symptom and requires careful consideration but in spite of previous writings there are few neurologists who would make a diagnosis of increased intracranial pressure on an analysis of headache alone and the following must be taken only as generalizations It may be frontal lateral occipital or diffuse it may be described as throbbing bursting or by other adjectives but probably none of these has any notable significance The duration is more important if a patient seeks advice for a headache which has been present only for days weeks or even months and is more or less continuous increased intracranial pressure must be suspected Such a headache is usually inconstant as regards severity and tends to be affected by posture frequently it is worse after lying down and improves after standing being present on waking and becoming less severe during the morning At the same time the headache may disappear altogether if the patient remains in bed particularly if he is propped up The pain may be aggravated by coughing sneezing and defaecation as well as by a lumbar puncture It will always be temporarily relieved to some extent by even the simplest of analgesics such as aspirin or phenacetin

If a group of patients complaining of headache is analyzed the number suffering from increased intracranial pressure will be found to be small personal experience has shown that intra cranial tumour accounted for less than one in fifty When the symptom is of long standing almost half of the cases will be found to have purely psychogenic headache almost a third will have the recurrent headache of migraine and most of the rest will have had a head injury apart from these three conditions chronic headache is rarely seen Psychogenic headache tends to be described in extravagant terms as regards severity and is often bizarre in distribution being commonly vertical or localized

as bands or linear strips it is often unaffected by any analgesics

Psychogenic headache may be worse on waking—when the problems of life have again to be faced In the neurotic subject headache is never an isolated symptom and the patient always has other complaints insomnia being especially frequent Headache however may be the only symptom of increased intracranial pressure Unilateral or localized headache has little relationship to the site of a brain tumour but there is certainly a correlation between pain referred to the occipito cervical region and tumours in the posterior fossa

The mechanism by which *vomiting* is produced in intracranial hypertension is not certain It is in fact uncommon and very rarely a presenting symptom It may or may not be associated with nausea and may present explosively but an intracranial cause should always be remembered as a possible explanation of simple recurrent vomiting Relatively rare as a presenting symptom of tumour it is almost constant in the symptomatology of subarachnoid haemorrhage Vomiting ultimately appears in about fifteen per cent of patients with cerebral tumour and when profuse it is often an index of impending disaster

Papilloedema is a particularly important finding because it can assume considerable proportions before the patient makes any complaint of visual disturbance Since the fundus is unlikely to be examined in a patient who has no symptoms papilloedema is rarely a presenting manifestation and is in fact not often discovered as an early physical sign though it becomes increasingly common in those with progressive symptoms The ophthalmoscope is the physician s most important and completely irreplaceable instrument but unfortunately there are few members of our profession apart from the ophthalmologist and those addicted to neurology who are able to recognize papilloedema especially in its early stages It has little localizing or lateralizing significance even though it may at first be unilateral It should be and usually is possible to diagnose or at least suspect early papilloedema before haemorrhages have made their appearance on the basis of a disc that is pinker than normal with less clearly defined borders abolition of the physiological pit engorgement of the retinal veins and swelling (a three dimensional visual observation) especially of the nasal edge There are many causes

other than increased intracranial pressure and indeed the commonest of all is vascular (malignant) hypertension Al though the finding of papilloedema is of the greatest importance its absence does not contraindicate the diagnosis of cerebral tumour indeed it frequently does not appear at all especially with slowly growing tumours which infiltrate the brain without adding much to the intracranial contents such as the more chronic forms of glioma (astrocytoma and oligodendroglioma) The duration of the history is not of importance in this context. Papilloedema can appear very quickly as a result of intracranial thrombophlebitis or haemorrhage but it is often absent in subdural haematoma or brain abscess with a history of similar duration Papilloedema is eventually followed by failing visual acuity constricting visual fields and sluggish reaction of the pupils to light Finally if the increased pressure is not relieved papilloedema (which may have been quite unsuspected) will be followed by irreversible optic atrophy with blindness however in the early stages severe depression of vision cannot be attributed to papilloedema alone Sudden and severe unilateral loss of vision with papilloedema often occurs in the retro bulbar neuritis of disseminated sclerosis and rarely results from pressure on the optic nerve by a cerebral aneurysm Although very much less common than headache in the syndrome resulting from increased intracranial pressure papilloedema may exist without headache or vomiting and it ultimately appears in about two fifths of patients with cerebral tumour When intracranial pressure is high the victim frequently complains of dizziness or unsteadiness but this is not an early symptom More specific attacks of vertigo may have important and early localizing significance and will be discussed later

EPILEPSY

In a personal survey epilepsy has proved to be the commonest presenting symptom of a brain tumour though only slightly more frequent than headache and it ultimately occurred in half of these patients It is much more likely to appear early than late in the illness being in this regard similar to headache and unlike papilloedema the incidence of which increases with the passage of time Fits which are associated with a space-

ocupying lesion may be general (major) or focal The former which are commoner have no specific features but if they are preceded by an aura this may have important localizing significance Focal fits take many forms The classical focal motor (Jacksonian) attack may start in hand face or foot and spread fairly quickly say from fingers to face and then perhaps to the leg but sometimes it remains strictly confined to one limb There is no disturbance of consciousness but after the attack the affected limb may be weak or even paralysed for an hour or more Such an episode suggests that the focus of origin is in or near the cerebral cortex of the opposite side in front of the fissure of Rolando Sometimes the attack spreads to the opposite side of the body when consciousness will be lost but the motor aura has the same focal significance as when it is an isolated incident A similar spreading sensory aura with various sensations including numbness tingling or even pain suggests a cortical point of onset posterior to the fissure of Rolando Temporal lobe seizures include a wide variety of phenomena One of the first to be identified was the uncinate fit with its olfactory hallucination and dreamy disturbance of consciousness there may also be gustatory visual or vertiginous features Temporal lobe seizures also include the episodes formerly labelled as psychomotor epilepsy and in a previous era epileptic or psychic equivalents In these there is a phase lasting two or three minutes in which purposive or semi purposive and repetitive movements are carried out in a state of disturbed consciousness with subsequent complete amnesia During such an attack the patient is unlikely to fall or to be incontinent but lip smacking and fiddling with the fingers are frequent Temporal lobe seizures are one of the commonest of all epileptic manifestations and are often misinterpreted as hysterical behaviour

Although epilepsy is the commonest presenting symptom of a cerebral tumour the converse is not true It used to be stated that the likeliest cause of epileptic attacks first appearing in adult life was a brain tumour but few would now agree with this and in many such patients the aetiology of the epilepsy remains obscure even after the most exhaustive investigation Certain pathological processes are particularly epileptogenic **Cerebral cysticercosis** is always associated with epilepsy and is

an uncommon but important cause of fits appearing in adult life occasionally it closely simulates the symptomatology of a single space occupying lesion **Cerebral angioma,** which is usually not a tumour but a developmental anomaly has a particular tendency to produce epilepsy a history of fits with a previous attack of **subarachnoid haemorrhage** is almost diagnostic though the diagnosis is nowadays always confirmed or refuted by cerebral angiography **Brain abscess** when supratentorial is often accompanied by fits which are frequently the presenting symptom

FOCAL SYNDROMES

Focal syndromes appear in at least half the patients with cerebral tumour and perhaps the commonest is a progressive hemiplegia which is the presenting symptom of one in ten cases Occasionally *hemiplegia* of sudden onset may be produced by focal brain ischaemia resulting from an unsuspected tumour In the elderly or hypertensive subject such a tumour is likely to remain unrecognized until headache papilloedema or other evidence of expansion appears but the possibility should always be borne in mind When sudden hemiplegia appears for no clear reason in a young person with a normal blood pressure and without valvular heart disease or other obvious source of embolism a space occupying lesion should be suspected Such a sudden and sometimes completely reversible hemiplegia is not uncommon as evidence of a cerebral angioma but it may also be the presenting symptom of a glioma A slowly progressive hemiplegia is not always an expression of a space occupying lesion and indeed it is now known to be one mode of presentation of internal carotid artery occlusion. With tumours involving the frontal temporal or parietal lobes of the dominant hemisphere (the left in right handed people but not always the right in the left handed) some form of *dysphasia* is likely to appear not infrequently as the presenting symptom and it may easily be overlooked As with hemiplegia an attack of aphasia may be sudden and in the first place transient One of the commonest early speech defects is nominal dysphasia—inability to name objects or persons in the early stages the victim will often mask

his disability by circumlocution and the difficulty will be made apparent only by asking him to name common objects or better still to enumerate four animals, birds flowers or trees When dysphasia is predominantly of the sensory or receptive kind speech is often so disturbed and incoherent that the patient is regarded as psychotic and in the absence of other abnormal findings on physical examination he may well be admitted to a mental hospital

A focal sign of great importance frequently undetected because the patient is unaware of his disability is *homonymous hemianopia* which constitutes about fifteen per cent of the presenting focal syndromes Such a visual disturbance will never be missed if it is looked for and simple testing by confrontation can be carried out in a few seconds Homonymous hemianopia or homonymous quadrantic defect which is as easy of recognition may result from a lesion anywhere between the optic chiasma and the occipital cortex though it is most commonly associated with masses in the temporal region It may then be accompanied by dysphasia hemiplegia and sensory loss

Lesions of the parietal lobe may produce sensory disturbance of all kinds in the opposite limbs but an important and sometimes solitary phenomenon is that known as *sensory suppression* or *extinction* which is also easy of demonstration if the possibility is kept in mind In this condition a stimulus (pain touch or pressure) is appreciated on the affected side of the body but if an identical stimulus is applied simultaneously to the same point on the opposite limb the patient will be aware only of the stimulus on the normal side sensation on the affected side being suppressed

Although tumours of the posterior fossa become less common with advancing years they may be seen at all ages Neurological examination and history taking are of necessity limited in young children Cerebellar tumour in infancy (usually medulloblastoma or astrocytoma) may produce a vague syndrome of listlessness or fretfulness for weeks or months with *unsteadiness of gait* Cerebellar signs may or may not be easily elicited but tend to be most marked in the legs nystagmus is inconstant and bulbar signs unusual

About a third of all posterior fossa tumours and almost ten per cent of the neurosurgeon s material are acoustic neuromata The important presenting sign here is *unilateral deafness* of which the patient may make no complaint Headache is common as an early symptom and amongst early signs are ipsilateral cerebellar ataxia loss of ipsilateral corneal reflex (which is occasionally associated with a complaint of facial numbness) nystagmus (which is usually present) and sometimes attacks of rotatory vertigo though the latter may also result *from purely cerebellar tumours* Acoustic neuroma should be diagnosed before the appearance of papilloedema and vomiting in the presence of which operative mortality is appreciably raised

Cranial nerve palsies are common and important but some times misleading signs The most frequent a sixth nerve palsy *is often found in a patient with increased intracranial pressure* from any cause because the long intracranial course of this slender nerve renders it particularly liable to damage from stretching However such a palsy sometimes has localizing value *Unilateral anosmia* will only be discovered if sought and is an important manifestation of an ipsilateral tumour especially a meningioma (liable to grow in the olfactory groove) or a frontal glioma Because of direct pressure a frontal tumour may result in ipsilateral anosmia and failing vision culminating in increas ing pallor of the optic disc while as the tumour increases in size the opposite disc may develop papilloedema—a diagnostic syndrome

When *multiple cranial nerves* are involved in a progressive syndrome in which there is no involvement of speech nor any disturbance in the limbs and especially if the palsies are uni lateral the probable explanation must always be a tumour involving the base of the skull Such a picture is likely to have one of three explanations **Nasopharyngeal carcinoma** is not uncommon and presents either by involvement of lymph nodes in the neck by unilateral deafness from occlusion of the Eustachian tube or by infiltration of the base of the skull with involvement of several cranial nerves on the same side the diagnosis may ultimately be established only by examination of the nasopharynx under general anaesthesia A similar involvement of the base of the skull with cranial nerve palsies usually

on one side but sometimes on both may be caused by **metastases** from any malignant growth especially from bronchial and mammary carcinoma The precise diagnosis is now of increasing importance because of the outstanding therapeutic results of adrenalectomy in metastatic mammary carcinoma—even when the primary lesion may have been treated as many as twenty years previously Less commonly a syndrome of multiple cranial nerve involvement is the result of a **meningioma,** which is then likely to be inoperable

MENTAL CHANGES

With increasing intracranial pressure the patient usually becomes lethargic apathetic and drowsy but such symptoms are late or terminal Sometimes however a cerebral tumour may present purely as a problem of slowly developing personality change with or without features of dementia Psychotic symptoms are perhaps maximal when a glioma appears in one hemisphere and begins to involve the corpus callosum but tumours arising in one frontal lobe may be particularly silent as regards physical signs of an unequivocal nature When a patient has had epileptic attacks for weeks months or even many years and then develops progressive changes in conduct and personality even in the absence of physical signs a frontal tumour should be suspected this is particularly important because of the possibility of successful surgical amputation of the affected frontal lobe Frontal tumours may result in urinary incontinence of unusual type the patient micturating irrespective of time or place and with a total disregard of social implications—mental incontinence In the presence of vascular hypertension the differential diagnosis of dementia may be extremely difficult and there is no specific psychotic picture which is peculiar to cerebral tumour

SPECIAL PATHOLOGICAL CONDITIONS

Many different pathological processes involve the **pituitary gland** or hypophyseal stalk The commonest of these is the pituitary chromophobe adenoma of adults which expands the sella

turcica and usually presents by virtue of pressure on the optic chiasma resulting in a defect in the temporal visual fields This defect starts as a paracentral scotoma but by the time the patient is first seen there is often a complete bitemporal hemianopia easily detectable on confrontation At this stage there may be evidence of hypopituitarism including such symptoms as obesity amenorrhoea impotence lack of the need to shave and a falling blood pressure Such tumours are eminently operable but also frequently responsive to radiotherapy A chromophil adenoma results in gigantism or acromegaly according to age but such skeletal changes may be unassociated with radiological evidence of enlargement of the sella or involvement of the visual path ways these tumours may also respond well to radiotherapy

Two pathological processes which mimic cerebral tumour and often present with focal symptomatology are brain abscess and subdural haematoma

Brain abscess is most commonly otogenic and is then almost invariably found in the temporal lobe or cerebellar hemisphere complicating either acute or chronic otitis media on the same side The clinical picture is one of headache sometimes with vomiting and contralateral hemiparesis with or without dysphasia or quadrantic visual field defect Cerebellar abscess is charac terized by ipsilateral limb ataxia Whatever the situation the patient is usually apathetic and drowsy and it is important to remember that papilloedema pyrexia and leucocytosis are often absent in any case the two latter are of dubious diagnostic significance in the presence of acute otitis Metastatic abscess from chronic pulmonary suppuration may be found anywhere in the brain but an abscess complicating sinusitis (especially the frontal sinus) is likely to be adjacent to the source of infection The incidence of this condition has fallen since the advent of sulphonamides and antibiotics though the mortality is still considerable

Chronic subdural haematoma is a collection of blood between the dura and arachnoid membranes and is therefore intracranial and intradural but extracerebral It results from damage to small veins allowing a slow escape of blood leading to the development of a cyst containing fluid blood or clots Although trauma is the main factor in its production the amount of the

initial damage is often trivial and in about half the cases there is no clear evidence of injury After an interval usually of days but occasionally of weeks or even months the patient develops increasing headache with or without vomiting papilloedema or weakness of limbs on one or both sides subdural haematoma is usually parasagittal but quite often bilateral The appearance of drowsiness ptosis a dilated and fixed pupil ocular palsy or slowing of the pulse rate call for urgent surgical intervention Subdural haematoma is nearly always diagnosed on clinical evidence alone but is occasionally an unexpected discovery in a patient suspected of a cerebral tumour It constitutes one of the most satisfactory of neurosurgical problems though it is fatal if unrecognized

Conclusions

Early diagnosis is always important even in conditions which though recognized are nevertheless untreatable The mortality from cerebral tumour remains very high but the pituitary (chromophobe) adenoma the subdural haematoma and many meningiomata offer opportunities for complete and permanent surgical relief Much can be done for the acoustic neuroma and the brain abscess and gratifying results sometimes follow surgical extirpation with or without radiotherapy of the more benign forms of glioma Even solitary metastatic tumours have been removed with success and increasing knowledge of steroid chemistry makes the early diagnosis of secondary growths important and potentially rewarding

CHAPTER XIII

POLIOMYELITIS

By Michael Kremer

POLIOMYELITIS is an acute infective disease due to the spread in the body of a specific virus which may invade the nervous system When it does it attacks mainly the anterior horn cells of the spinal grey matter and the motor cells of the brain stem nuclei but this is usually preceded by a phase of irritation of the meninges producing a characteristic meningeal reaction with spasm in the muscles of the back and neck

Until 1947 the disease occurred only sporadically in this country but since that year considerable epidemics have occurred though they have never risen to such heights as in the United States During epidemic periods the diagnosis is made very readily as always in any epidemic but the sporadic case often presents considerable difficulty When the condition is atypical the diagnosis is frequently suspected without definite proof being achieved

To understand the manifestations some knowledge of the epidemiology and etiology is necessary as well as of the spread within the body and this will be very briefly considered It has been shown that whenever poliomyelitis occurs there is a good deal of minor illness in the vicinity and that there may be as many as a hundred or more instances of minor illness for each case of frank poliomyelitis Usually in a household in which a case of paralytic poliomyelitis occurs almost every member has been infected and virus may be isolated from them all though the occurrence of several paralytic cases in one household is rare The virus may be isolated both from sewage and from flies in the neighbourhood thus indicating a very easy mode of spread

There are three types of virus now clearly isolated These are the Brunhilde the Lansing and the Leon (types 1 2 and 3) with different morbidity features The virus can be obtained from the nasopharynx of acute cases of healthy contacts of convalescents and also from the stools of the same groups Some small epidemics have been traced back to a healthy individual who has been in contact with a paralytic case though never developing the disease himself The usual route of infection is through the alimentary canal and then to the central nervous system either through the blood stream or the autonomic nerves from the alimentary canal Probably the former is the more usual route Exceptionally the pharynx or tonsils may be the portal of entry—particularly the exposed raw area after tonsillectomy Inoculations even months earlier (as with pertussis vaccine) may precipitate the disease but are more likely to mark the region which will be most severely affected

There is now no doubt that muscular effort or trauma of any kind in the early stages of the disorder increase the chances of a much more severe paralytic phase This is true in general but also true for individual muscles and groups of muscles and it stresses the need for complete rest and gentleness of handling in these early stages It also calls during epidemic periods for the abandonment of injections (particularly intramuscular ones) and the postponement in susceptible groups of all but the more important operations

Once the infection has entered the body complete rest is at present the only means of influencing the course of the disorder the need for early diagnosis is therefore vital Unfortunately except in epidemics or where there is a newspaper scare the doctor is often not asked to see the patient until these early stages have passed when it may be found that the patient has tried to work off the trouble —a hazardous process to be condemned out of hand

Early diagnosis is also important socially in attempting to limit the infection by some degree of isolation but as already indicated this may be impossible by the time the diagnosis is made because the virus is already so widely distributed Amongst small communities however the spread may take place through such narrow channels that efficient isolation of one such com

munity may be very effective in preventing the others from becoming involved

MANIFESTATIONS OF POLIOMYELITIC INFECTION

There are now known to be four ways in which the body may react to the poliomyelitis virus

The commonest is the development of *immunity without symptoms of any kind of illness* This probably occurs in the majority of infected people and study of the occurrence of immune bodies in the blood indicates that full protection is present by the age of forty five in most of us

The next largest group is that of *minor illness* in which general symptoms are present without the nervous system being involved These are the abortive cases

The third group comprises those in whom the nervous system is involved without the anterior horn cells being attacked There are changes in the cerebrospinal fluid which indicate that invasion of the nervous system has taken place but immunity builds up so rapidly or the virulence is so low that paralysis never occurs These are therefore called *non paralytic*

The fourth group which is the smallest comprises the *paralytic* cases

The second group is probably impossible to diagnose except in the middle of an epidemic since the symptoms are identical with those of almost any general infection There may be nothing but malaise headache and some pyrexia lasting a few hours This may clear up rapidly or continue and become more severe until signs of meningeal irritation appear

In the preparalytic stage it may be possible to separate two clinical phases or they may merge into one another The first symptoms are fever of rarely more than 101 together with malaise and headache Sometimes there is also drowsiness alternating with restlessness or insomnia flushing sore throat anorexia nausea vomiting and diarrhoea There may be a non specific backache and pain on movement of the eyes This phase may last two or three days and may then be followed by improve ment for a similar or briefer period

In the first stage the signs are few The patient is obviously ill the headache is typically the generalized vascular headache of fever which is made worse by coughing or straining or jarring The flushed face congested conjunctivae and inflamed fauces are exactly the same as those found in any virus infection during the stage of viraemia

The second stage marks the invasion of the nervous system The temperature rises again though it is rarely very high being usually below 103 and most frequently between 100 and 101 The headache becomes more severe and in addition to the generalized headache previously present pain is now present in the occipital and suboccipital regions spreading down the back with a complaint of stiffness in the affected muscles Pains of a deep aching character are present in the limbs together with hyperaesthesia of both skin and muscles Often fleeting paraesthesiae are complained of and in young children con vulsion may occur at any stage The gastro intestinal symptoms may return at this time and vertigo may be present In the more severe forms confusion amounting to delirium may supervene but this is uncommon

Examination now will show a much more ill patient with mucosal congestion as before who is more difficult to examine because the muscles and skin are so tender There is frequently some resistance of variable degree to flexion of the neck together with a diminution in straight leg raising (Kernig s sign) The back muscles may be the only ones affected and passive flexing of the spine may be impossible In a child this may be shown by a failure to kiss the knees Spasm and tremor in muscles may be felt and a fine nystagmus is frequently present at this stage Formal examination of the nervous system may reveal the nystagmus but rarely shows any change in power or reflexes it is questionable in view of the deleterious influence of effort whether detailed testing of the limbs for power is justified In non paralytic cases the patient recovers after these manifestations have been present for a few days

The onset of paralysis usually comes rapidly after the pre paralytic phase and is associated with severe pains in the limbs and an increase in the tenderness of the muscles on pressure It is rare for the preparalytic phase to last longer than a few

days but sometimes it continues for one or even two weeks Occasionally muscle fasciculation or intense spasm may precede the paralysis It is impossible to forecast the severity of the paralysis from the severity of the preparalytic phase but it is usual for the most tender muscles to show the greatest paralysis The weakness is always patchy and asymmetrical unless the bulk of the muscles are affected Usually the maximum amount of damage is done in the first twenty four hours and a careful watch must be kept on the patient without more than the minimum of formal testing of the muscles In a few cases the weakness may occur in bursts when it is usually found that a rise in temperature heralds each fresh step in the paralysis some times the paralysis is steadily and rapidly progressive and then takes the form of an acute ascending or descending palsy It is always rash to say that the chance of further paralysis is over until the temperature has been normal for a week for rarely the infection may smoulder on for this time and then a few more muscles may suffer Fortunately the extent of the acute paralysis is usually much greater than the ultimate permanent weakness

Examination in the early paralytic stage shows weakness and hypotonia though the latter may be masked by spasm in other muscles or even in parts of an incompletely paralysed muscle Muscle tenderness may persist for a long time The tendon reflexes are usually diminished or may be absent if the respond ing muscles are involved Neck and back stiffness and diminished straight leg raising may also persist for some weeks

The early stages of involvement of the respiratory muscles must be recognized for there is no doubt that anoxia increases the severity of the paralysis in all muscles Probably the simplest means is to *train the patient to count at a fixed rate after taking the deepest breath possible* A fall in the count indicates a decrease in the vital capacity and pulmonary ventilation In hospital spirometers are used for this purpose but this requires even greater training of an ill patient and needs skilled personnel to ensure reliable instrumental results To wait for obvious failure in intercostals and diaphragm and for the appearance of cyanosis is dangerous both to life and to the recovery of paralysed muscles

Involvement of the brain stem appears to be increasing in frequency and affects the cranial motor nerves from the trigeminal downwards The eye muscles are rarely involved It is very important to recognize these cases early since there is difficulty in swallowing and coughing which leads to an accumulation of saliva in the back of the throat and of mucus in the respiratory passages The risk of inhalation of fluid is great and respiratory embarrassment is early The postural drainage of accumulated material and its clearance by a sucker is life saving in this condition It is clearly vital to distinguish anoxia produced by palsy of the muscles of respiration from that produced by accumulation of saliva and mucus in the airway for the treat ment is very different In the former assistance to the weak muscles is given early by a negative pressure respirator of the box type In the latter postural drainage and suction may be enough but if there is any associated weakness of the respiratory muscles then assistance can only be given with safety by per forming a tracheotomy and using a positive pressure pump through a cuffed tracheotomy tube

It is clear that the management of respiratory failure of any kind in poliomyelitis calls for a special centre with highly trained nurses and doctors but the early recognition of the threat of respiratory failure is vitally important because postural drainage can be carried out anywhere and this may give time for a mobile respiratory resuscitation team to be summoned from such a special centre

INVESTIGATIONS

The only investigations usually carried out in the early stages of the disease are examination of the blood and spinal fluid The former usually shows little change except for a possible mild lymphocytosis or monocytosis In the early stages the spinal fluid may show no changes except for a minor rise of pressure but as the stage of meningeal irritation occurs the pressure rises and a pleocytosis is produced The cells may number anything from fifty to 500 per ml made up equally of polymorphonuclear cells and lymphocytes but after a week only lymphocytes are found Protein is only slightly raised at first with an increase in

globulin but goes on rising for several weeks reaching a figure of 300 mg to 500 mg per 100 ml Chloride and sugar content are usually normal The investigation of respiratory function by measurement of ventilation and blood gas content is beyond the scope of this chapter as is the study of the virology of the disease

DIAGNOSIS

As has already been indicated this is rarely possible in the stage of general invasion except during an epidemic Then the tendency is to label any constitutional disturbance as polio myelitis Only the isolation of the virus from the individual followed by a subsequent rise in the specific immune bodies to that virus in the blood could prove the nature of such a constitu tional disturbance and this is clearly impossible The diagnosis therefore at this stage is from any viraemia and is only made in retrospect

In the stage of meningeal involvement examination of the spinal fluid will help but the condition must be differentiated from other causes of meningeal irritation Pyogenic meningitis usually shows greater pyrexia and the patient looks more ill in the early stages while the meningeal irritation may be greater The spinal fluid shows a greater and purely polymorphonuclear pleocytosis often with the causative organism present The sugar is low or even completely absent The chlorides are not affected

Tuberculous meningitis may present difficulty but the pro dromal period is often longer and the degree of asthenia is often great The spinal fluid may show both polymorphs and lympho cytes with an excess of protein and both the sugar and chloride are diminished Tubercle bacilli may be found

Lymphocytic meningitis may be found in association with mumps glandular fever or chorio lymphocytic virus infection but the general manifestations and blood changes usually help the differentiation

Acute febrile polyneuritis may have much the same premoni tory symptoms while the signs at this stage which include tender muscles spasm and paraesthesiae may be indistinguishable Soon definite sensory loss may be found of a typically peripheral

distribution Sometimes the polyneuritis may appear to be purely motor on clinical examination and the differentiation then becomes very difficult Polyneuritis should be suspected however if the motor loss is remarkably symmetrical and involves the distal parts of the limbs most severely in the early stages Spinal fluid examination may help since in polyneuritis the cellular content is little if at all increased while the protein content is high from the first It is most important to differentiate the conditions as corticosteroid therapy may have a profound effect in polyneuritis

In young children where there is a good deal of pain and spasm such general conditions as acute rheumatism must be considered as well as epiphysitis and osteomyelitis but these are usually far more localized in their tenderness and joint involvement is characteristic of the first two It is possible that in adults acute spinal lesions such as thrombomyelia transverse myelitis or the sudden enlargement of a spinal cyst may be confused but in any of these the flaccidity below the lesion found in the acute stage is associated with extensor plantar responses and sensory loss Fairly soon the flaccidity may diminish and some tone return even to the point of spasticity

In recent years epidemics of a peculiar encephalomyelopathy have occurred often associated with myalgia and some enlargement of lymph glands These have been seen best in semi closed communities such as nurses homes The Middlesex Hospital and Royal Free Hospital epidemics were typical and such cases have been reported from many parts of the world Before the epidemic is established differential diagnosis is difficult because the general symptoms may be similar to those of poliomyelitis and signs of meningeal irritation may be present together with severe pain in the limbs and great tenderness of skin and muscles Peripheral muscular weakness may be present but there are also often signs of brain stem and cerebral hemisphere involvement Sensory changes may be present and the finding of a normal spinal fluid helps to establish the diagnosis

The bulbar form of poliomyelitis must be distinguished from other forms of encephalitis Lethargic encephalitis is now very rare and the presence of external ophthalmoplegias with or without pupillary changes will differentiate the condition En

cephalitis following exanthemata or vaccination should present no difficulty because of its time relation to the primary condition

At one time it was taught that an attack of poliomyelitis protected the individual completely but there are now many well authenticated cases of multiple attacks of poliomyelitis The isolation of the different groups of poliomyelitis virus makes the reason for this quite clear for an attack can confer immunity against the attacking virus alone

The large scale vaccination of susceptible individuals against the three types of poliomyelitis virus is now being carried out in many parts of the world and it is still too early to say how much protection will be afforded by this measure At present it is suggested that about seventy per cent protection will be produced but this may have to be modified with increasing experience What must be clear however is that alterations in the clinical manifestations will occur and it may well be that when the bulk of the population has been protected the whole of this chapter on early diagnosis of poliomyelitis will need to be rewritten

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CHAPTER XIV

DISSEMINATED SCLEROSIS AND PARKINSONISM

BY HENRY MILLER

DISSEMINATED or Multiple Sclerosis and Parkinsonism are the two commonest structural diseases of the nervous system in Britain at the present time and the average general practice of six thousand patients includes three or four sufferers from each disorder In both instances diagnosis is often long delayed It must be admitted at once that neither condition offers an encouraging field for therapy In the case of disseminated sclerosis there is for practical purposes no effective treatment, beyond the management of symptoms and an attempt to adjust the patient psychologically to her disability As for the medical treatment of Parkinsonism the continually augmented profusion of drugs advertised is adequate testimony as to their very limited effectiveness Surgical measures amongst which electrical coagulation of the globus pallidus attracts current interest will probably never be applicable to more than a small minority of Parkinsonian patients and they are in any case still in the phase of experimental development Under such circumstances and despite the relative frequency of these diseases it might be thought that their prompt diagnosis would be a matter of little practical importance but this is not so It is remarkable for example how often the victim of Parkinsonism has to make his own diagnosis often in the face of his doctor s scepticism and sometimes after labouring for a long period under the unjustified imputation of psychoneurosis It may be that he loses little by some delay in discovering the serious and progressive nature of his illness but this is rarely his own reaction to the situation and it is indeed especially in the earlier and often protracted stages of the disease that considerable subjective disability may be effectively controlled by treatment It is possible in the case

of disseminated sclerosis to adduce certain arguments against early diagnosis and there are certainly good reasons to justify withholding such information from the sufferer Yet even in this disease a firm diagnosis is not infrequently welcomed with positive relief by a patient who has felt the burden of months or years of diagnostic doubt and confusion sometimes with more than a suggestion of medical evasiveness There are also very cogent reasons positively in favour of early and definitive diagnosis which implies in the case of this disease more than the mere relief of exoneration from a suspicion of neurosis or of apprehension about some even more serious possibility All too often for example the diagnostic confusion which arises from failure to elicit or to appreciate the case history of early disseminated sclerosis has led to fruitless and unnecessary investigations to myelography ventriculography or worse Again while this disease may at present be untreatable accurate differential *diagnosis remains vital to exclude conditions with which it may* easily be confused some of which such as spinal tumour or subacute combined degeneration of the spinal cord are eminently responsive to prompt treatment Furthermore at any time some unexpected therapeutic advance may endow the early recognition of the disease itself with an importance as critical as that which now attaches to the once entirely academic diagnostic problem of early tuberculous meningitis Finally since in the present state of knowledge a diagnosis of disseminated sclerosis tends to absolve the doctor from further investigation or therapeutic effort it is clearly at least as important to make such diagnosis definitive as to make it early

These two diseases are similar in that their recognition is essentially a problem of clinical medicine depending on the informed interpretation of a careful history and examination ancillary methods have little to offer in the matter

DISSEMINATED SCLEROSIS

The early symptoms of disseminated sclerosis are clinical manifestations of the characteristic pathological lesions of the disease which are plaques of acute demyelination occurring in successive showers and scattered throughout the white matter of

the central nervous system Some such lesions remain clinically silent but a proportion whether because of their intensity their extent or their critical situation interfere sufficiently with conduction through a group of nerve fibres to produce a disturbance of physiological function which reveals itself as a neurological symptom or sign These lesions are so widely distributed that almost any focal neurological symptom may occur in disseminated sclerosis and it is indeed curious that there are a few such as hemianopia aphasia and flaccid palsy with loss of deep reflexes which are rarely encountered However since there is such a vast range of possible and recorded presenting symptoms it might be anticipated that early diagnosis would present insuperable difficulties That such difficulties are in fact less frequent than might be expected is due to the fact that while no individual symptom is particular to disseminated sclerosis the combination or pattern of symptoms and their natural history are often immediately revealing This is indeed an instance where a diagnostic method working from general principles is more reliable as well as more intelligent than one based on an endeavour to identify text book types of disease entity

Briefly the key to the accurate diagnosis of disseminated sclerosis is implicit in the statement that while it may be suspected it cannot be firmly sustained unless there is evidence of **dissemination of lesions both in space and in time** For example one of the most characteristic symptoms which this disease may present is an *acute retrobulbar neuritis* usually though not invariably unilateral Dimness of vision in the affected eye is often followed by severe visual failure and is usually associated with pain in and above the eye and tenderness of the globe on pressure The condition in most instances develops over the course of a few days and clears up during the ensuing weeks leaving an often imperceptible patch of blindness near the fixation point and some pallor of the optic disc Disseminated sclerosis is much the commonest cause of this syndrome but there are other occasional causes Retrobulbar neuritis may arise as an apparently allergic sequel to a variety of general infections especially of a virus nature It is an occasional complication of local orbital or sinus infection and a very rare manifestation of herpes zoster and of endarteritis in the secondary stage of syphilis The attribu

tion of such a lesion to disseminated sclerosis depends on evidence of involvement of other parts of the nervous system in the previous history or on neurological examination If examination of a patient presenting with retrobulbar neuritis discloses nystag mus an extensor plantar response or asymmetrical abdominal reflexes the presence of a coincident lesion elsewhere in the nervous system is evident (dissemination in space) and a strong suspicion of disseminated sclerosis arises If dissemination in time is also evident in a previous history of transient neurological symptoms such as diplopia a clumsy hand or a dragging foot suspicion ripens into certainty

No decade is entirely immune from the onset of disseminated sclerosis and the writer has seen it in typical form at seven and a half and in the sixties but three quarters of all cases begin between the ages of fifteen and thirty five It accounts indeed for so large a proportion of cases of structural nervous disease in early adult life that it is usually the first such diagnosis to be considered in this age group

Weakness of one or more limbs due to a demyelinating lesion involving the pyramidal tract in the spinal cord is the commonest initial symptom and the disease presents in this way in nearly half of all cases The lower limbs are involved more often than the upper and such involvement is often asymmetrical and insidious the disability being first noticed during a long walk or while hurrying The leg or legs are described as feeling stiff weak or heavy Examination at this stage will usually reveal increased knee and ankle jerks with an extensor plantar response on one or both sides and asymmetry rapid fatigability or total absence of abdominal responses Very frequently in such cases the symptoms may be entirely unilateral but the physical signs indicate bilateral involvement of the pyramidal tracts These are of course non specific signs of disease involving the lateral columns of the spinal cord and the same considerations relevant to multiplicity of lesions apply as in the account of retrobulbar neuritis given above In this instance the diagnosis may be clinched by the finding of a pathologically pale optic disc or of nystagmus or by unearthing a past history of transient diplopia some years previously Occasional cases occur especially in middle age in which an insidiously progressive entirely un

remittent and sometimes symmetrical lesion of the pyramidal tracts in the spinal cord usually also with some degree of sensory impairment has arisen as the only manifestation of disseminated sclerosis the nature of the disease being subsequently confirmed at autopsy Such cases are included in the small minority of instances of this disease in which it may be necessary to employ every ancillary method of investigation to exclude spinal tumour or some other surgically treatable cause and in which diagnosis may not only be impossible of early achievement but must necessarily remain provisional for many years

A less common initial symptom than weakness of a lower limb and sharing second place in order of frequency with retro bulbar neuritis is *subjective disturbance of sensation* (par aesthesiae) The commonest cause of such a symptom is a plaque of demyelination involving the posterior columns of the spinal cord most often in the cervical but sometimes in the dorsal region The abnormal sensations are usually most marked peri pherally in the hands or feet and may be unilateral or bilateral sometimes spreading to the trunk The complaint may be of numbness tingling pins and needles electric shocks a feeling of cold water running down the limb or of a tight string around the extremity Often paraesthesiae can be provoked in all four limbs by forward flexion of the neck Sometimes the affected part may feel either continually constricted or swollen beyond its usual size Under any of these circumstances cursory examina tion of the limb may reveal little or nothing amiss but the descriptions and analogies employed are so stereotyped that they clearly describe physically determined disturbances of sensation and are never to be dismissed as emotional symptoms Indeed careful examination in such cases will practically always disclose some definite impairment of discriminative sensation such as vibration or positional sense appreciation of passive movement or two point discrimination in the affected part For this reason there is often a complaint of clumsiness of the affected limb The hand is useless and cannot carry out fine movements or appreciate the nature and texture of objects placed in it or the patient feels a lack of control over the affected leg

Amongst other frequent modes of onset in disseminated sclerosis *diplopia* due to a mid brain lesion and *acute vertigo*

caused by a plaque disturbing vestibular connection in the pons deserve special mention The diplopia of disseminated sclerosis may be of brief duration lasting for only a day or two on the other hand a complaint of merely momentary double vision is of no clinical significance Strabismus like ptosis is relatively rare and coincident weakness of conjugate ocular movement or actual nystagmus often bespeak the central origin of the lesion This disease is the commonest cause of transient double vision in early life just as arteriosclerosis is responsible for most cases of this syndrome over the age of fifty Vertigo is often abrupt in onset severe and accompanied by gross ataxia and vomiting The condition is usually to be distinguished from Meniere s syndrome both by the relative youth of the patient and by the absence of deafness or tinnitus Furthermore the vertigo often persists for several days and less severely even for a matter of weeks If examination reveals other evidence of the disease such as optic atrophy reflex changes or impaired vibration sense diagnosis is of course easy

Some less common clinical manifestations are important *Bladder symptoms* more often occur late than early but occa sionally herald the onset Frequency urgency and hesitancy are often accompanied by some constipation and nearly always by reflex changes below the waist Disseminated sclerosis should always be suspected when typical *trigeminal neuralgia* affects a young subject it may become bilateral *Facial paralysis* in distinguishable from Bell s palsy is remarkable as the only common instance of a lesion of the lower motor neurone in this condition A few cases begin with an apparently *cerebellar* picture but the classical signs of nystagmus ataxia scanning speech intention tremor and titubation of the head are essen tially those of the advanced disease Quite a proportion of cases diagnosed as disseminated sclerosis of cerebellar type are suffer ing in fact from insidious cerebellar degeneration with an entirely different prognosis

The symptoms and signs described above indicate some of the anatomical sites in which the ubiquitous plaques of this disease are especially prone to produce symptomatic disturbances It must be stressed again that the enumeration of such signs is of less importance for diagnosis than a clear appreciation of the

general symptom pattern which emphasizes the remittent and multiple nature of the characteristic lesions Also of great practical importance in diagnosis are the *psychological aspects* of the disease A few patients with disseminated sclerosis react to the disability by a predominantly depressive mood not in any way incongruous with their predicament Rather more become inordinately labile during the course of the disease fluctuating between extremes of cheerfulness and gloom with excessive and uninhibited emotional reactions The often recorded occurrence in this disorder of schizophrenia as of tuberculosis is in all probability nothing more than the coincidence of diseases with a similar age incidence while dementia when it occurs is usually late However probably more than half of all patients with this disease exhibit *euphoria* at some stage of the disorder often as an early sign and often throughout the protracted course of their illness The importance of this one of the commonest single clinical features of the disorder can hardly be over estimated The contrast between serious disability and sometimes almost fatuous cheerfulness often associated also with a quite abnormal sense of physical well being is a unique clinical phenomenon Euphoria is rarely encountered in the neurological disorders which are commonly confused with disseminated sclerosis and although it is valuable only as a positive sign every experienced neurologist can recall cases in which it has clinched an otherwise equivocal diagnosis

Although spinal puncture in patients with this disorder may undoubtedly be followed by a constitutional upset or even on rare occasions by an actual exacerbation of the disease the frequency of such events has been greatly exaggerated and the vast majority of patients with disseminated sclerosis tolerate the procedure without disturbance Nevertheless spinal fluid examina tion is superfluous in the majority and essential only in clinically doubtful cases Its greatest value is in excluding the possibility that a paraplegic syndrome may be due to compression of the spinal cord by tumour or some such lesion Less often it may provide confirmatory evidence that some minor neurological disorder is due to early disseminated sclerosis while in any case a grossly abnormal fluid renders the diagnosis untenable How ever except for the finding in a minority of instances of a paretic

Lange curve with negative serological tests for syphilis the very slight changes found in disseminated sclerosis are non specific and the fluid is of course often entirely normal

Differential Diagnosis

Since the signs of the disease are so numerous and so variable differential diagnosis covers potentially the whole of neurology However some practical difficulties are encountered often enough to deserve special mention Early disseminated sclerosis is often wrongly diagnosed as **hysteria.** There are many reasons for the notorious difficulties which arise in this connection The organic significance of early subjective sensory symptoms may not be appreciated especially when examination reveals no unequivocal abnormality Or a frankly hysterical complaint such as a bizarre gait may respond promptly to suggestion and persuasion but may nonetheless conceal a substratum of early organic disease In some instances hysterical exaggeration and elaboration appears to be a way of drawing the doctor s attention to a physical dis ability which the patient feels to be insufficiently appreciated Furthermore the euphoria of disseminated sclerosis may be mistaken for the heroic fortitude and cheerful indifference typical of the hysterical reaction Hysteria is in any case one of the most difficult diagnoses in medicine to sustain with confidence as Osler said it deceives even the very elect It should certainly never be made unless there are positive psychiatric as well as negative physical findings Even so mistakes are inevitable it is not unknown for episodes of conversion hysteria to herald the onset of disseminated sclerosis which declares its lf in abnormal physical findings only on repeated examination over a period of months

The frequently remittent course of **syringomyelia** may cause confusion In this condition however careful anatomical diag nosis will indicate an essentially local lesion in the cervical cord or brainstem the central situation of which is evident in persistent superficial loss of sensation to painful and thermal stimuli This is scarcely ever encountered in such dense or dissociated form in disseminated sclerosis where any such findings are usually both partial and fugitive in this disorder the characteristic sensory loss is of posterior column type (vibration sense positional

sense tactile discrimination) rather than due to lesions of the spino thalamic tract When disseminated sclerosis remains limited to the spinal cord compression by **tumour** or a **disc lesion** must be rigorously excluded by spinal fluid examination and myelo graphy and the same applies in many middle aged patients in whom damage to the spinal cord by the multiple disc lesions and osteophytosis of **cervical spondylosis** (osteo arthritis) may mimic the disease In older patients also gastric analysis blood examination marrow puncture and vitamin B_{12} assay may be necessary to exclude **subacute combined degeneration** of the spinal cord which may present in an atypical form and in which both optic atrophy and mental changes may occasionally be encountered

When disseminated sclerosis presents initially with an acute episode of severe neurological illness its differentiation from the **acute disseminated encephalomyelitis** which may complicate non specific infections such as those of the upper respiratory tract may present great difficulty In general fever pain meningism and loss of deep reflexes are rare even in the most acute episodes of the chronic disease but any of them may occur In such cases the unspoken reservations which the physician feels about his confident and reassuring diagnosis of an acute self limiting disease may be all too fully confirmed during subsequent months or years of observation

PARKINSONISM

This condition is unique in clinical neurology in that its recognition is to a much greater extent a matter of total and immediate impression than of careful history and meticulous physical examination It has indeed been truly said that unless the disease is recognized as the patient enters the consulting room its diagnosis may defy the most searching investigation The writer has seen a patient lying in a hospital bed under specialist observation as a case of progressive hemiparesis attributed to a probable tumour of the cerebral hemisphere (but with a para doxically flexor plantar response!) in whom a more experienced observer entering the ward was able to make a confident and immediate diagnosis of Parkinson s disease on the basis of a casual glance

Even an experienced physician may sometimes find prolonged observation necessary for definitive diagnosis many ex service men have continued to draw pensions for neurasthenic syndromes supposedly of emotional origin during the years of periodic neurological re examination which were needed gradually and finally to confirm an early suspicion of organic striatal disease Conversely there are other patients in whom the early signs of Parkinson s disease may be clearly recognizable long before there is any hint of relevant symptoms

Parkinsonism is in essence nothing more than the non specific syndrome of damage to the corpus striatum So much is clear from its rare occurrence after head injury and following carbon monoxide or manganese poisoning while it may also very occasionally be caused by primary or secondary tumour involving the base of the brain Furthermore its features often form a part of other and more generalized syndromes such as cerebral arteriosclerosis traumatic encephalopathy (punch drunk) or hepatolenticular degeneration However such occasional examples are numerically insignificant in comparison with the large majority of cases of Parkinson s disease *per se* Thirty years ago most subjects of this disease could be confidently allotted to one of two fairly clear cut groups—post encephalitic Parkinsonism or paralysis agitans The first of these affected patients under the age of forty rigidity was usually more striking than tremor while associated ocular autonomic and psychological symptoms were frequent Paralysis agitans lacked these last features and had a maximal incidence between the ages of fifty and sixty tremor being more conspicuous than rigidity Today the virtual disappearance of acute encephalitis lethargica has altered the situation and the majority of current cases of Parkinson s disease present a more indeterminate picture

Parkinsonism is today a disease of middle or late middle age and the patient in an advanced stage of the disorder is easily recognizable even by a layman because of his flexed posture forward tottering gait muscular immobility asymmetrical tremor expressionless face and monotonous voice Early in the course of the disease however diagnosis is often extremely difficult In a few patients indeed the condition presents as nothing more than a generalized weakness with retardation of all movements

It is particularly in these cases that an initial diagnosis of neurasthenia tends to be made since systematic examination may yield no abnormal findings In most such instances however some of the minimal signs described below will be found if they are carefully sought In the majority of patients there is in any case some localization of symptoms In general it may be said that initially these are usually *unilateral* that they affect the *upper limb* more than the lower and that *rigidity* is commonly more conspicuous than tremor The greatest difficulties arise in the occasional such case where tremor appears to be entirely lacking In some patients a deterioration in handwriting may be the first symptom this tends to become smaller and the letters to decrease in size along the line but if tremor is pronounced this may be clearly evident in the script In other cases decreasing agility at the piano or a rattling teacup may first attract attention

An abnormality of *gait* may be the earliest sign Long before a gross change has developed it may be observed that the patient is not swinging his right arm freely as he walks The sign next noticed is most often *facial immobility* The face which is always bilaterally involved lacks the normal play of expression it may with difficulty be provoked to unfreeze into a slow and hesitant smile and it then immediately resumes its inexpressive stare Indeed perhaps the most consistent finding disclosed by inspection is *infrequent blinking* which is almost pathognomic of Parkinsonism and it is not at all uncommon for a firm diagnosis to be possible on the basis of this sign alone As he sits in the consulting room the patient s *general immobility* is striking He never fidgets and does not move from time to time to re adjust his position in the chair but sits impassive and unmoving his hands crossed in his lap or where tremor is embarrassing concealed under his coat or in his pocket The *tremor* of Parkinsonism again affects the arm more than the leg and usually begins and remains more severe on the right side It is so stereotyped in its pattern and so familiar to every doctor that it is indeed remarkable that it should in its early stages often be misinterpreted as hysterical The main reason for this—and also the reason why the disease itself was for so long generally regarded as a neurosis—is its variability In its beginnings it is always intermittent It disappears during sleep and at rest but begins anew

and often increases markedly in amplitude as soon as attention is directed to it Some doctors evidently still find it difficult to believe that such behaviour can be compatible with an organic origin

The further physical signs revealed by systematic examination in the early case are often scanty and only two groups are of real importance Examination of ocular movements will usually reveal a marked impairment of convergence though this is of course in any case common after middle age Sometimes there is rapid fluttering of the closed lids though like oculogyric crises a greasy face and excessive salivation this was a more striking feature of frankly post encephalitic cases than it is of the common indeterminate examples of Parkinsonism seen today The cardinal and pathognomic ocular sign however is what has been described as the *glabellar tap* sign When the bridge of the nose is rhythmically and gently percussed with the finger in the normal subject synchronous blinking continues for a few seconds and then gradually ceases In Parkinsonism blinking on percussion of the bridge of the nose will continue regularly in response to such percussion for as long as this repeated stimulus is applied The other important physical finding on neurological examination is rigidity of the limbs This is most easily elicited in the affected arm In the clasp knife spasticity of a pyramidal tract lesion such as is seen for example in the hemiplegia which follows a cerebral thrombosis stiffness of the arm yields abruptly and completely to firm passive flexion after an initial phase of involuntary resistance In disease of the corpus striatum however rigidity yields slowly and uniformly to sustained pressure in a manner perfectly described by the traditional analogy of the lead pipe Cases with a marked element of tremor manifest phasic interruptions in an otherwise similar plastic rigidity giving the equally familiar and characteristic impression of a cog wheel

The differential diagnosis of Parkinsonism is less complicated than that of many other neurological disorders Indeed it is a disease of which it can be said that to think of it is to diagnose it Parkinson s disease is overlooked usually because the possibility has never crossed the doctor's mind though he recognizes its presence immediately it is pointed out to him The same

curious phenomenon occurs with a few other disorders amongst which myasthenia gravis myxoedema and polyarteritis nodosa immediately suggest themselves

One of the conditions with which Parkinsonism is sometimes confused is cerebral arteriosclerosis Indeed Parkinsonian features may occur in such a case but they are invariably complicated by evidences of a more widespread lesion The patient with arteriosclerotic Parkinsonism deteriorates by a series of downward steps (ischaemic incidents) rather than insidiously There may indeed be a frank clinical history of successive little strokes or an extensor plantar response on one or both sides and almost invariably there is some concomitant evidence of failure of concentration or of actual intellectual deterioration Such findings are of course entirely lacking in purely striatal disease Even the sluggishness and croaking voice of hypothyroidism or the staring eyes of its converse have been known to cause confusion but as with the forced immobility of the old man with severe myocardial insufficiency such difficulties should surely be little more than momentary A much more difficult problem may be presented however by the elderly patient with a severe retarded depression Striatal disease itself leading as it does to a progressive constriction of the patient s horizon is usually accompanied by a depressive outlook of greater or lesser degree and of course suicide is not unknown in this condition However in a few cases of depressive retardation where the co existence of Parkinsonism has been suspected the rapid resolution by electroconvulsive treatment of immobility muscular rigidity and an attitude of flexed misery indicates their occasional origin on the basis of a functional rather than a structural lesion

CHAPTER XV

FITS AND FAINTS

By J B Stanton

THE term fits and faints used in the title of this chapter is designedly imprecise It is intended to refer to attacks of sudden loss of consciousness which may or may not be accompanied by other features such as change of colour muscular twitchings convulsive movements and incontinence of urine and which are usually short lived and tend to recur It is not intended to include single attacks of loss of consciousness with an obvious precipitating cause such as sudden loss of blood cerebrovascular accident or head injury nor to refer to convulsions occurring in the course of uraemic states hepatic coma and eclampsia or during severe intracranial infections such as encephalitis or meningitis

It might appear at first sight that if during such recurrent attacks of loss of consciousness the patient has a convulsion then he is suffering from epilepsy and if he does not then he is suffering from syncope Unfortunately the position is not so simple as this since in some forms of epilepsy no convulsion occurs and on the other hand if syncope from whatever cause is prolonged the resulting cerebral ischaemia may give rise to a convulsion For this reason and because it is used in different senses by different authors the term syncope is avoided here A useful term to describe the attacks referred to in this chapter is that *most frequently used by the patients themselves namely blackouts* and it is the early diagnosis of these which is now considered

A patient may come or be brought for advice after a single such episode but more commonly he has had several attacks before medical opinion is sought The practical importance of early diagnosis of such attacks is abundantly clear Not only do

curious phenomenon occurs with a few other disorders amongst which myasthenia gravis myxoedema and polyarteritis nodosa immediately suggest themselves

One of the conditions with which Parkinsonism is sometimes confused is cerebral arteriosclerosis Indeed Parkinsonian features may occur in such a case but they are invariably complicated by evidences of a more widespread lesion The patient with arteriosclerotic Parkinsonism deteriorates by a series of downward steps (ischaemic incidents) rather than insidiously There may indeed be a frank clinical history of successive little strokes or an extensor plantar response on one or both sides and almost invariably there is some concomitant evidence of failure of concentration or of actual intellectual deterioration Such findings are of course entirely lacking in purely striatal disease Even the sluggishness and croaking voice of hypothyroidism or the staring eyes of its converse have been known to cause confusion but as with the forced immobility of the old man with severe myocardial insufficiency such difficulties should surely be little more than momentary A much more difficult problem may be presented however by the elderly patient with a severe retarded depression Striatal disease itself leading as it does to a progressive constriction of the patient s horizon is usually accompanied by a depressive outlook of greater or lesser degree and of course suicide is not unknown in this condition However in a few cases of depressive retardation where the co existence of Parkinsonism has been suspected the rapid resolution by electroconvulsive treatment of immobility muscular rigidity and an attitude of flexed misery indicates their occasional origin on the basis of a functional rather than a structural lesion

hysterical and anxiety attacks Examples of the second group include disturbances secondary to local cerebral vasospasm in hypertensive encephalopathy to disturbances of cardiac rhythm such as the *bradycardia of heart block to the effects of irritation* of the carotid sinus to postural hypotension and to organic heart disease together with emotional fainting Attacks of loss of consciousness due to alterations in the composition of the blood may occur in tetany from whatever cause and in hypo glycaemic states Certain toxic substances such as alcohol cocaine and the organic arsenicals may affect the metabolism of the brain sufficiently to cause convulsions these may also follow the abrupt withdrawal of long continued barbiturate medi cation

A wide range of disorders may precipitate attacks of un consciousness through the mechanisms just described and an important step in early diagnosis is the distinction between those attacks which are the results of a purely psychogenic disorder those which result from a temporary disturbance of function of the nervous or cardiovascular systems and those which are a symptom of some established underlying disease Some of the more important causes in these categories are tabulated below but no attempt at completeness is implied

1 Psychogenic hysteria
2 Temporary and recurrent functional disturbances idiopathic epilepsy (*i e* without demonstrable organic disease) cardiac dysrhythmias fainting and postural hypotension irritability of the carotid sinus tetany (hypocalcaemic and alkalotic) and hypoglycaemia
3 Organic diseases epilepsy resulting from congenital *traumatic neoplastic infective degenerative and vascular* disease of the brain hypertensive cerebral attacks heart block (Stokes Adams attacks) aortic stenosis and aortic insufficiency and toxic causes

The differential diagnosis of these conditions in their early stages poses a number of problems and many of the difficulties arise from the fact that it is unusual for the doctor actually to witness the attack himself More often he has to rely on the account (necessarily incomplete) given by the patient and if these are available on descriptions of varying value given by

they cause considerable alarm to the sufferer who wishes to know first and foremost whether they are likely to recur but they also raise important considerations with regard to the employment and the social acceptability of the patient himself For instance if the attacks are diagnosed firmly as epileptic the patient may well have to change his employment and rearrange his life particularly if he is engaged in an occupation which necessitates work at heights or near moving machinery and other hazards of this type or which requires constant alertness such as driving on the road or railways The shifting fashions in social attitudes to these attacks of loss of consciousness may also have their influence upon the patient s everyday life There is ample evidence of the mixed horror and fascination with which the falling sickness has been popularly regarded through the ages and little Matilda s epileptic fits were more acceptable in the Victorian drawing room when they were referred to as her faints in spite of the damp patches left on the carpet Similarly there can be no doubt that the vapours common in young ladies of the upper classes during the nineteenth century were regarded at that time with more patience and sympathy than such manifestations of hysteria or affectation would arouse today Early diagnosis may make possible a favourable prognosis or an effective regime of treatment which will relieve the patient from these and similar anxieties

The problem in early diagnosis of blackouts is less one of quibbling whether the patient has had a fit or faint than one of detecting the underlying causative condition and pursuing the implications of this for prognosis and treatment It is useful to consider first of all the mechanisms by which such recurrent attacks of loss of consciousness may be brought about These may be divided into two groups First those in which there is primarily **an episodic disturbance of function of the nervous system,** either due to purely psychogenic causes as in hysteria or accompanied by paroxysmal disorder of cerebral activity as in epilepsy Secondly those in which there is primarily a **disturbance of the blood supply to the brain,** due either to temporary disturbance of the circulation or to variations in the composition of the blood As examples of blackouts produced by the first mechanism we may instance cases of epilepsy of all sorts and

on maintaining the erect position Attacks due to epilepsy hypoglycaemia cerebral vasospasm and heart block are likely to be independent of posture If the patient is standing up at the onset of an epileptic attack he will in many cases fall down immediately but this is not the case in petit mal or psychomotor epilepsy By contrast the patient who faints is likely to sway at first, and then sink more slowly to the ground

Thirdly enquiry should be made for the occurrence of any *associated symptoms* When convulsions have occurred it is important to learn whether these were focal unilateral or generalized at the onset since a focal origin is evidence of epilepsy A report of any change in the patient s colour such as pallor or cyanosis may be of value in indicating the possible mechanism responsible for the attack Incontinence of urine and biting of the tongue often accompany an epileptic fit, but do not occur in ordinary fainting When tetany is the cause of the attack palpitations a feeling of numbness in the extremities and around the mouth or twitching of the muscles may be experienced before any loss of consciousness

Fourthly the *duration of the seizure* may give some clue as to its nature If the length of the attack was very brief lasting only from a few seconds to a few minutes then petit mal epilepsy Stokes Adams attacks fainting and postural hypotension are the most likely causes A duration of more than a few minutes but less than an hour is suggestive of hypoglycaemia or grand mal epilepsy

The *length of the history* may act as a diagnostic guide in some cases For instance attacks which have recurred over a period of years and which first made their appearance in childhood or adolescence are most likely to be the result of idiopathic epilepsy or epilepsy secondary to birth trauma After careful questioning about the events surrounding the attacks themselves it will be necessary to enquire into the general health of the patient and into such relevant points in the past history as the occurrence of birth *injury or subsequent head trauma meningeal or encephalitic* illnesses childhood convulsions rheumatic fever and nephritis Any evidence of epilepsy in the patient s relatives may be stoutly denied at first but the possibility should always be carefully explored When the history has been obtained a full

witnesses of the attack In most cases the diagnosis has to be based on the history including the description of the attacks themselves and on the physical examination of the patient although as will be mentioned later certain ancillary investigations may sometimes be of help When the history is being taken it is important that careful attention should be given to the circumstances leading up to the attack and to any factors which may have appeared to influence it When the patient and witnesses have told the story in their own words valuable information may often be elicited by direct questioning on the following points

First the *mode of onset* of the attack should be determined in particular whether this was sudden or gradual and whether the patient had any warning since this information may provide valuable diagnostic clues For instance the occurrence of a recognizable brief aura at the onset suggests epilepsy If the attack had an instantaneous onset epilepsy or disturbance of cardiac rhythm are the most likely causes If however the onset lasted over a period of a few seconds accompanied by a feeling of dizziness then postural hypotension fainting or irritation of the carotid sinus are more likely Often an emotionally disturbing event may be shown to have preceded a fainting attack or a hysterical fit Hypoglycaemic attacks usually have an onset over a period of several minutes during which the patient may become mentally confused sweat copiously and turn pale

Secondly it is important to establish *what the patient was doing* at the time of the attack whether he was standing sitting or lying down at the onset and whether he fell in the attack An attack which disturbs sleep is practically always epileptic Attacks associated with lowering of the blood pressure including carotid sinus attacks usually occur only in the sitting or standing positions and those resulting from postural hypotension are apt to occur immediately after a change from the recumbent to the erect posture Prolonged standing in military parades or sitting in a warm room often precede a faint and an onset during or immediately after exertion suggests the possibility of aortic stenosis attacks following exertion are likewise occasionally seen in patients with aortic regurgitation Paroxysmal tachycardia as a rule leads to unconsciousness only if the patient insists

causes of fits and faints included in the third group listed above In many of these cases further examination by special techniques will be required before the diagnosis can be firmly established and treatment can be planned If such an examination does not reveal the presence of abnormal physical signs between the attacks then the differential diagnosis will rest between those conditions included in the first and second groups described Conscious malingering or the simulation of epileptic fits is very rare and in practice the distinction usually lies between epilepsy in all its forms fainting and hysteria Features which may enable a distinction to be made between these three conditions will therefore be described first followed by an account of some more exotic but not uncommon causes of sudden loss of consciousness

Epilepsy may occur in various clinical forms The diagnosis is not difficult when the attack consists of a typical grand mal convulsion In this case the patient loses consciousness suddenly often with a loud involuntary cry and falls immediately to the ground A stage of tonic spasm ensues with increasing cyanosis and is followed in fifteen to thirty seconds by clonic movements of limbs and trunk together with incontinence of urine Focal epilepsy can readily be recognized by convulsive movements starting in one hand or foot or at the corner of the mouth which subsequently spread by a characteristic march to involve neighbouring parts and sometimes to become generalized A similar pattern occurs in focal sensory attacks In petit mal epilepsy which is seen most typically in children the patient does not usually fall but he may suddenly become pale and lose consciousness for a few seconds only tonic spasm and convulsions do not appear but there may occasionally be rhythmic blinking of the eyes and twitching of the face In yet other forms of epilepsy particularly those associated with disturbances of the temporal lobe and in cases of grand mal inadequately controlled by medication the patient may fall to the ground unconscious without any convulsive movements and a period of disordered behaviour constituting one form of psychomotor epilepsy may follow such an attack Difficulty often arises in differentiating this type of attack from fainting but certain distinguishing features may give an indication of the

clinical examination of the patient must be undertaken including estimation of the blood pressure and examination of the urine The examination should be directed particularly to eliciting any signs which may provide evidence of anaemia or of organic dis turbance in the cardiovascular and central nervous systems

If the patient is by good fortune seen in an attack then attention should be paid to those features which may indicate the nature of the mechanism involved The character of the breathing the colour of the skin the appearance of the veins and the heart rate may all be of considerable diagnostic value When the attack is due to disturbances of the peripheral circula tion such as emotional fainting and postural hypotension pallor may be striking but it is not accompanied by cyanosis or respiratory disturbances and the veins are usually collapsed If the attack is the result of diminished cerebral circulation conse quent upon cardiac disorder there is likely to be a combination of pallor and cyanosis with distension of the veins and *dyspnoea* In epilepsy initial pallor or flushing may be followed by cyanosis and venous congestion during the early stages of the convulsion Any evidence of convulsive movements during the period of un consciousness will naturally suggest epilepsy but it must be remembered that prolonged cerebral anoxia and hypoglycaemia can also lead to convulsions The limitation of such convulsive *movements at the onset of the attack to one part or to one* side of the body alone will be a more valuable indication suggesting epilepsy of focal origin Examination of the state of the reflexes is often of value in distinguishing hysterical from other types of fit, for in the former the reflexes are all normal and indeed considerable resistance may be encountered when attempts are made to elicit the corneal responses On the other hand in major epilepsy and in states of prolonged or deep loss of consciousness from whatever cause the corneal and pharyngeal reflexes are often absent the tendon reflexes are sluggish and the plantar responses may be extensor In cases of heart block the striking *bradycardia of thirty per minute or less can be observed* and in hypoglycaemic attacks the pallor and sweating of the patient will be prominent features

Careful physical examination of the patient between the attacks will usually reveal the presence of any of the organic

Postural hypotension results from a similar mechanism to that which operates in fainting namely an inadequacy of reflex vaso motor control It may occur in elderly subjects who have lost weight and whose muscles have become flabby through being confined to bed and also after the operation of lumbar sym *pathectomy or in patients receiving ganglion blocking drugs* for the treatment of hypertension Postural hypotension may occa sionally be seen as a result of similar autonomic denervation in organic disease of the nervous system especially in tabes dorsalis Attacks of loss of consciousness due to postural hypotension can be recognized by their clear cut relationship to change of posture and measurement of the blood pressure changes between the recumbent and erect positions will confirm the diagnosis

The fact that a patient has had a definite convulsion during an attack of loss of consciousness does not permit an immediate diagnosis of epilepsy since similar convulsions may occur to wards the end of a prolonged attack of cerebral ischaemia from circulatory causes or during the course of hypoglycaemic attacks Convulsions may also appear during hysterical attacks but these are becoming more and more rare With the advent of the Age of the Common Man the exuberant displays of Charcot s *grandes hysteriques have given place to less colour* ful performances more consonant with the exigencies of our time **Hysterical fits,** when they are seen nowadays occur mostly in women of low intelligence who in addition often show evidence of trauma to the brain at birth The differentiation between hysterical and epileptic fits becomes apparent when the history is taken Hysterical attacks are clearly related to situa tional stresses they do not occur in the absence of an audience and in spite of their apparent violence the patient seldom injures herself If the attack is witnessed the struggling but co ordinated *movements of the limbs with screaming and shouting the absence* of incontinence and the resistance to examination together with the normal state of the reflexes and plantar responses remove any possible doubt about diagnosis The fact that a patient has had an hysterical fit and presents other symptoms of hysteria should not however be taken to exclude the diagnosis of epilepsy since the two conditions may and frequently do occur together In many cases an initial epileptic fit may be followed by hysterical

correct diagnosis and of the different physiological mechanisms involved in the two cases

Fainting occurs when the blood pressure in the cerebral arteries falls with resulting cerebral anoxia Such anoxia may be the result of anaemia and of organic disease of the heart such as heart block aortic regurgitation or aortic stenosis Far more commonly how ever fainting occurs in the absence of any such organic basis and in these cases it is a result of instability of vasomotor control which leads to pooling of the blood in the muscles of the limbs and in the viscera with a consequent diminution of venous return to the heart. Fainting usually occurs as a sequel of obvious external influences such as pain the sight of blood disturbing emotional experiences or prolonged standing to attention in military parades Fainting may also occur in a hot room even when there are no precipitat ing emotional causes Fainting does not occur when the patient is in the recumbent position and indeed his recovery is hastened by lying down Epileptic attacks on the other hand occur in the absence of precipitating emotional factors and they may occur when the patient is lying in bed In epilepsy conscious ness is lost more suddenly than in fainting Feelings of dizziness and light headedness may precede fainting but are of little diagnostic significance since they may also be experienced in epileptic seizures On the other hand the presence of a recogniz able epileptic aura is of considerable value in diagnosis The aura commonly takes the form of an unpleasant and indescribable feeling in the epigastrium which seems to rise up to the head at which point the patient becomes unconscious It may also consist of an hallucination of the sense of smell taste sight hearing or touch or it may manifest itself by sudden unexplained emotional changes such as feelings of depression fear or anger A sudden return of normal consciousness at the end of the attack is in favour of a diagnosis of epilepsy for the patient who has fainted usually recovers more slowly and often has a feeling of physical weakness Similarly if the patient exhibits drowsiness or mental confusion for some time after recovery of conscious ness if he complains of headache or if he vomits the attack was most probably epileptic while the occurrence of automatism during this period is proof of its epileptic nature

found fall in the prolonged blood sugar curve Reactive hypo glycaemia which occurs three to four hours after a large carbo hydrate meal in persons of faulty dietary habits does not as a rule lead to actual loss of consciousness or convulsions

Attacks of unconsciousness resulting from **irritability of the carotid sinus** occur typically in elderly arteriosclerotic patients Pressure on the carotid sinus in a normal person will cause bradycardia and a fall in blood pressure However in a patient with hypersensitivity of the carotid sinus even light pressure such as is caused by turning the head so that a tight collar presses on the neck is likely to cause an attack of sudden loss of consciousness particularly if the patient is standing or sitting upright at the time

Another occasional cause of attacks of loss of consciousness is sometimes known as '**cough syncope**' In this condition which is seen most frequently in victims of chronic bronchitis and emphysema the extreme rise of intrathoracic pressure to wards the end of a paroxysm of coughing diminishes the venous return to the heart and precipitates unconsciousness and even convulsions In these cases a clear cut history can be obtained of an attack occurring at the end of a prolonged bout of coughing In some cases unconsciousness appears to accompany a single cough and not a paroxysm but in these patients the mechanism is different and the cough should probably be regarded as an integral part of an epileptic fit arising from focal cortical irritation

Elderly patients and especially women of menopausal age occasionally suffer from sudden falls which are referred to as '**drop attacks**', and these may be a source of considerable embarrassment particularly when they occur in the street These attacks are so unexpected and short lived that it is often impossible to determine whether the patient has really lost con sciousness They usually take the form of a sudden fall on to the knees for which the patient can find no apparent reason and from which she immediately recovers Such attacks usually tend to disappear over the course of a few years The mechanism of drop attacks is not known and there is no conclusive evidence that they are epileptic since they do not respond to anti

manifestations on recovery and this is particularly the case in epilepsy arising from a focus in the temporal lobe If the initiating fit has been of a minor type it may even have passed unnoticed by the onlookers An important guide in these cases is a history of definite epileptic fits (major or minor) in the past which were *not* followed by hysterical behaviour Another way in which hysteria may bring about ' fits or faints is through the mechanism of tetany This is a consequence of hysterical over breathing which causes carbon dioxide deficiency and leads to muscular twitching and carpopedal spasm Both in hyper ventilation tetany and in hysterical fits the patient may claim to be unconscious during the attack but she will nearly always admit she was aware of what was going on although unable to speak or to control her movements Attacks of dizziness short of actual loss of consciousness are not uncommon in patients suffering from anxiety neurosis

We may turn now to some more unusual causes of fits and faints which are nevertheless seen with sufficient frequency to warrant a discussion of their differential diagnosis The effects of **tetany** caused by hysterical over breathing have already been mentioned but tetany may also arise as the result of prolonged vomiting or of the ingestion of large amounts of alkalis for the relief of dyspepsia Tetany under these conditions is due to alkalosis without diminution of the serum calcium It may also however accompany a fall of the serum calcium level in such conditions as hypoparathyroidism following thyroidectomy rickets and osteomalacia The occurrence of carpopedal spasm during the attacks and evidence of latent tetany such as a positive Chvostek s sign will help to establish the diagnosis Another cause of obscure attacks of unconsciousness is **hypoglycaemia,** either as the result of an overdose of insulin in a known diabetic or occurring spontaneously in cases of islet cell tumour of the pancreas The latter cause may be suspected when the fits occur after a period of fasting typically in the early morning when the patient first arises The hypoglycaemic seizures are commonly attended by periods of disordered behaviour and progressive mental deterioration The final diagnosis of this condition will rest upon the demonstration of a low fasting blood sugar level and of the characteristic pro

Secondly recourse may be had to ancillary investigations These include X ray of the skull which may assist in cases of suspected intracranial disease the estimation of fasting blood sugar levels and the charting of blood sugar curves in patients suspected of pancreatic adenoma electrocardiography in patients suspected of intermittent heart block and the use of the electro encephalograph

Considerable faith is often placed in the electroencephalogram as a means of distinguishing between epilepsy and fainting but such faith is likely to prove ill founded unless the limitations of the method are realized The electroencephalogram of any patient will fall into one of three categories the tracing may be normal it may demonstrate a diagnostic abnormality or it may show a non specific abnormality The significance of these results must always be assessed in conjunction with the clinical history and examination of the patient If the electroencephalogram of a patient with obscure attacks of loss of consciousness provides positive evidence of epilepsy or of local or generalized structural disease of the brain associated with epilepsy then the diagnosis is made clear A normal record however is of less assistance since it does not rule out epilepsy (about one in three epileptics has a normal resting record) although it does render somewhat less probable the presence of gross organic brain disease as a cause of the attacks If the clinical findings give rise to a strong suspicion that a patient s attacks are epileptic a normal electro encephalogram should not be allowed to influence the diagnosis It is sometimes possible in such cases to produce a positive record by the techniques of activation of the electroencephalo gram These include making recordings after the patient has fasted for twelve to sixteen hours or after a period of several minutes voluntary over breathing or after administration of rapidly acting analeptics or barbiturate drugs Under any of these conditions latent epileptic features may become apparent in the tracing for the first time confirming the diagnosis If the electroencephalogram shows non specific abnormalities and the diagnosis from purely clinical data remains obscure it may be necessary to admit the patient to hospital for further investiga tion by means of lumbar puncture and the special techniques of

convulsant therapy and the electroencephalogram in such patients is normal

Patients with paroxysmal **vestibular disturbance** of sudden onset such as occurs in aural vertigo or Meniere s disease may occasionally be thrown to the ground but the history of severe vertigo at the onset the absence of convulsions the retention of consciousness during the attack and the presence of tinnitus and some diminution of hearing between the attacks will all serve to distinguish such episodes from those epileptic attacks in which an aura of giddiness is a prominent feature Occasional confusion may be caused by the occurrence of a vascular faint at the height of a severe attack of aural vertigo

When the detailed history and examination of a patient suffering from repeated attacks of loss of consciousness still leave doubt as to the probable cause of the attacks further investigations become necessary

First an attempt may be made to reproduce an attack Thus if a patient is suspected of having hypoglycaemic attacks these may sometimes be reproduced by giving an injection of insulin although patients with pancreatic tumours show a decreased sensitivity to insulin Carotid sinus sensitivity can be tested by pressure on one or other of the carotid sinuses but this is a procedure not without danger in arteriosclerotic patients and on no account should pressure be applied simultaneously to both carotid sinuses in such patients Postural hypotension may be observed by measuring the pulse rate and blood pressure while the patient changes from the recumbent to the erect position and hysterical hyperventilation attacks may be reproduced by inducing the patient to undertake prolonged voluntary hyperpnoea In all these tests it should be remembered that the procedures described frequently cause symptoms in a healthy person and the important point is not whether symptoms are produced but whether the test reproduces the exact pattern of symptoms which occurs in the patient s own spontaneous attacks The use of pitressin to bring about water retention and thus to precipitate fits in patients suspected of epilepsy has now been replaced by the various methods of activation of the electroencephalogram which are described below

CHAPTER XVI

COMA

By A G Ogilvie

THIS chapter will be devoted for the most part to the diag nosis at a reasonably early stage of a patient in coma rather than to the early diagnosis of coma itself Nevertheless it seems advisable even at the cost of some repetition to consider first of all the diagnostic features of incipient or imminent coma

Most commonly the onset of coma is sudden or so rapid that little or no warning of its imminence is given A maiden lady writing a letter is seized with a violent pain in the back of the head and falls forward on to the table unconscious an elderly man loses the use of his right hand and in less than half an hour is comatose Many patients pass from sleep into coma without waking In certain cases however premonitory signs and symptoms precede the onset of coma by many hours or even by a day or so giving some warning to the alert family doctor

The occurrence of nausea and vomiting persisting for twelve to twenty four hours in a known diabetic with or without pyrexia is an urgent indication for examination of the urine for the presence of sugar and ketone bodies If both are found in quantity the full treatment of diabetic coma is imperative even in a patient who is still conscious The observation of deep sighing respiration the so called air hunger of drowsiness and lethargy with increased thirst and depression of the deep reflexes are additional signs which offer valuable confirmatory evidence when they are present but it should be possible to recognize the imminent threat of diabetic coma in the absence of these classical features Furthermore it has to be realized that diabetes may present initially with coma and the above signs should encourage a suspicion of the diagnosis even in a patient not known to be a diabetic The occurrence of an acute infection or of a severe injury

cerebral angiography and pneumoencephalography in order to determine whether he harbours latent intracranial disease

In conclusion it may be emphasized that the commonest causes of recurrent attacks of loss of consciousness are **epilepsy**, whether idiopathic or symptomatic and **fainting of emotional origin** Other relatively common causes include **postural hypotension, hysterical hyperventilation,** and **carotid sinus sensitivity** **Spontaneous hypoglycaemia, paroxysmal tachycardia,** and **organic heart disease** are less frequent causes Fainting is often erroneously diagnosed in patients who are suffering from epilepsy Conversely there is a distinct danger that because of its frequency epilepsy may be mistakenly regarded as the cause of attacks of loss of consciousness which are in fact the result of postural hypotension or hypersensitivity of the carotid sinus Careful consideration of the features of the attacks and the circumstances in which they occur will enable this error to be avoided and will prevent the stigma (unfortunately still widespread) of epilepsy being unjustly attached to the patient

The ideal of early diagnosis is not merely to determine the nature of the attack—*i e* fit or faint —but to demonstrate the underlying mechanism involved and wherever possible its aetiology The diagnosis must be based upon a detailed history followed by full physical examination of the patient In obscure cases ancillary investigations such as X ray of the skull fasting blood sugar estimation and electroencephalography may be of some assistance but more often the diagnosis is revealed by painstaking enquiry into the circumstances surrounding the attacks It must be admitted however that in some cases only careful observation over a prolonged period of time will enable a firm diagnosis to be achieved Correct and early diagnosis of recurrent attacks of loss of consciousness will permit of a good prognosis in many cases and will enable effective treatment to be instituted in nearly all

of a reasonable degree of alveolar ventilation is the prime necessity

Three further instances of the so called pre comatose state may be mentioned An epileptic who is observed to sustain fits of increasing frequency and severity is a serious candidate for status epilepticus or perhaps epileptic coma and is best treated at this earlier stage Secondly the severely hypertensive patient who enters a phase of permanent and increasing headache together with a complaint of failure in mental and visual acuity must be regarded as a potential case of encephalopathic coma if not of a more serious catastrophe Finally an individual who suddenly experiences a severe headache followed by a period of unconsciousness stupor or mild confusion should be regarded as having sustained a slight leakage from an intracranial aneurysm and as a probable candidate for severe subarachnoid bleeding

In all these cases complete bed rest is immediately indicated in addition to the other remedies or procedures which are required in the particular case

CAUSES OF COMA

In the early diagnosis of the cause of coma it is necessary to bear in mind the range of possibilities The wider the doctor s vocabulary of causes the more readily and consistently will the diagnosis be made in practice Certainly the diagnosis which does not occur to him will not be made It is for this reason that the following summary is given here It is not complete and it is not intended to be complete Its purpose is simply to present a classification and to indicate the more usual causes of coma in each group It is anticipated that if the groups are considered the more unusual conditions will suggest themselves to the practitioner

1 **Local** (i e intracranial)

(a) *Vascular*	Intra cerebral haemorrhage Subarachnoid haemorrhage Cerebral thrombosis Cerebral embolism Cerebral oedema

in a diabetic patient should lead to the adoption of an actively expectant attitude quite apart from the other measures which are indicated in any case under such circumstances

If a known diabetic on insulin becomes restless and irritable sweats about the head and hands and feels faint and anxious he should be suspected of hypoglycaemia If he also has hallucinations of sight and hearing coma perhaps with convulsions due to a low level of sugar in the blood is imminent In a patient on soluble insulin early and prompt administration of glucose should prove an immediately effective remedy But in the case of a patient treated with one of the longer acting insulins rapid improvement may not take place In such a case and indeed in any patient who does not respond satisfactorily *before the doctor leaves the house* admission to hospital is necessary and urgent The clause in italics is emphasized because it is important that the doctor should wait until the effect or lack of effect of his dose of glucose can be observed If the doctor cannot wait he should return within an hour Patients are sometimes admitted to hospital in hypoglycaemic coma who have been given a dose of some form of sugar which has proved ineffective for its purpose either because it has been vomited or because it has been inadequate or because of continued insulin action Hypoglycaemic coma is dangerous because although death is uncommon serious sequelae may occur It must be remembered that severe and prolonged hypoglycaemia is equivalent to severe and prolonged anoxia and has the same effects Hemiplegia or mental disorder which may be temporary or permanent are amongst the more severe results but minor speech defects mental slowness and impairment of visual or auditory function also occur

A further important instance of this incipient phase is the chronic bronchitic patient with emphysema whose condition has recently and rapidly deteriorated usually due to an infective exacerbation If such a patient exhibits slight drowsiness with a tendency to mental confusion and perhaps twitching of the face and limbs he is liable to pass into a coma from which it will be extremely difficult to rescue him Treatment at this earlier stage is therefore urgently indicated The therapeutic problem is complicated and cannot be fully discussed here but restoration

This classification has been set out for the purpose of helping both the writer and the reader in the consideration of the subject under review There is no intention of allowing it to restrict unduly the scope and range of the discussion The diagnosis of the cause of coma will therefore be discussed first of all from the inside as it were in general conformity with the classified list just given and will then be considered again briefly from the outside in the diagnosis of a patient in coma

COMA OF 'LOCAL' OR CEREBRAL ORIGIN

This is for the most part the easiest section as the clinical *picture is usually straightforward although instances of extreme* difficulty occur from time to time

The vascular comas are probably the most familiar of all and little time need be spent upon them The rapid methodical progression of *intra cerebral haemorrhage in the hypertensive* subject is classical and is unmistakable if observed by or narrated to the doctor The *initial difficulty in speech* followed by clumsiness and weakness of an arm or of a leg and associated with progressive drowsiness stupor and coma cannot be overlooked The patient who is simply found unconscious may present some initial difficulty in recognition but the deepening unconsciousness the stertorous breathing which blows out the paralysed cheek and the flaccid tonelessness of the limbs on one side commonly make the position plain enough even for the relatives to recognize

The breathing in the case of cerebral haemorrhage does not greatly differ from that in coma due to massive thrombosis or embolism but apart from certain forms of poisoning and vascular accidents this stertorous type of noisy breathing is not observed in comatose persons and its occurrence narrows the diagnostic field Local paralysis manifest in loss of tone in a limb or limbs is not to be expected in cases of acute poisoning and its presence restricts the possibilities still further Thrombosis can be distinguished from haemorrhage only by an early or eventual return of consciousness though patients with thrombosis may in fact remain deeply unconscious for several weeks Fortunately the distinction is immaterial (except of course for

(b) *Injury*	Concussion Basal trauma Haemorrhage (intra cerebral subdural extra dural and subarachnoid)
(c) *Infection*	Meningitis Encephalitis
(d) *Space occupying lesions*	

2 **General**

(a) *Acute fevers*	Malaria typhoid fever etcetera	
(b) *Toxic*	The barbiturates carbon monoxide alcohol morphia etcetera	
(c) *Biochemical*	(i) Uraemia	Renal Alimentary Dehydration
	(ii) Endocrine	Diabetes Hypoglycaemia Adrenal and thyroid disorders Hypopituitarism
	(iii) Respiratory and circulatory	
	(iv) Cholaemia and portal systemic encephalo pathy	
	(v) Vitamin deficiency	
(d) *Epilepsy*		
(e) *Hysteria*		

Note —The possible reasons for administration or self administration of poisons have to be considered and a list of these is given below It should be borne in mind in taking the history —

Suicide	Abortion
Accident	Medical
Habit	Murder

Murder is a very uncommon cause of coma but it is in cluded because of the frequent complaint of judges about doctors in murder trials— they never think of poison It is the unusual that is most likely to be overlooked
The medical profession is directly involved in two ways It is perhaps not generally known that at least three or four patients annually are admitted in coma to any large general hospital due directly to the administration of morphia or hyoscine (and occasionally both) to a chronic bronchitic in respiratory acidosis or to a patient in status asthmaticus Other examples will no doubt come to mind Furthermore the practice of supplying a variety of drugs in tablet or capsule form in large amounts to patients of dubious emotional and mental stability has its dangers Such persons are liable to take just one extra for a really good night and so to pass into a state of semi automatism in which they automatically con tinue to swallow tablets until coma supervenes They are apt to be regarded as cases of suicide whereas they are simply victims of the urge to get as much as possible for the shilling The doctor is, of course only indirectly responsible here but the point is worth making

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believers in the anti coagulant treatment of cerebral thrombosis of whom the present writer is not one) The position is however different in the case of embolism in the young person who may be expected to have normal vessels In the writer s view such a patient requires intravenous Priscol and full anti coagulant therapy and diagnosis is therefore important The age the presence of a valvular lesion the history of sudden onset and the presence of local paralysis are the criteria and if these are satisfied no time should be lost By the time the patient reaches hospital it is often too late

The coma of intracranial tumour is often unrewarding as it is likely to be associated with the more malignant growths or with metastases from a bronchogenic cancer But its occurrence in cases of vertical and other meningiomata parasitic cysts established abscesses and so on renders a fatalistic attitude inexcusable A meningioma may be suspected if there has been a history of recurrent epileptic manifestations and there is usually some history suggesting intracranial disease But a negative history is of no value and indeed no history at all may be obtainable It is therefore necessary in all cases of unexplained coma to search for some local nervous sign and for evidence of increased intracranial pressure such as papilloedema The prognostic import is always grave but prompt attention to the urgent relief of intracranial tension may be successful particularly in the meningiomata

Cerebral oedema associated with hypertension (so called hypertensive encephalopathy) causes stupor and even unconsciousness but this is not usually very profound and the diagnosis is assisted by the presence of severe hypertension and often of retinitis and papilloedema It should be added that by severe hypertension is meant a diastolic pressure of at least 130 mm of mercury and a systolic pressure in accord with this Recognition implies also the absence of any other cause and the presence of hypertension alone is in itself insufficient as a basis for diagnosis

If the history is available as it usually is the diagnosis of subarachnoid haemorrhage should be readily made or the condition at least suspected The rapid onset of unconsciousness following sudden occipital headache is characteristic Occasionally however the patient may be comatose when first seen Rigidity

of the neck with or without (and usually without) Kernig s sign is to be expected and looked for The prognosis in subarachnoid haemorrhage with coma cannot be described as other than fair but recovery and even surgical cure are seen sufficiently often to make the early recognition and admission to hospital of such patients an urgent necessity The diagnosis can only be made by lumbar puncture and this must be done where doubt about the diagnosis such as the possibility of pyogenic meningitis exists Some caution is to be observed in the withdrawal of fluid from the spinal theca in these cases as bleeding may be encouraged by sudden alterations in cerebrospinal pressure but the diagnosis must be established The use of lumbar puncture as a diagnostic procedure in coma is probably undervalued It should be per formed in all cases in which the cause remains obscure after a full and careful clinical investigation

Consideration of the acute cerebrospinal infections which may now be conveniently discussed emphasizes this point The char acteristic history of onset of meningitis with severe headache and vomiting leading to unconsciousness is commonly obtained and the typical signs already mentioned are usually to be noted But on occasion the sole history is of drowsiness passing into coma or perhaps the illness begins simply with sudden un consciousness and no abnormal signs In any case of unexplained coma with fever therefore the possibility of acute septic meningitis should be excluded by lumbar puncture at a very early stage This early diagnosis is of particular importance in pneumo coccal meningitis where every hour counts and a delay of twelve hours may be fatal For though intensive systemic and intra thecal penicillin is curative it must be started within twenty four hours of the onset and preferably within twelve The writer does not refer to tuberculous meningitis as this must be diagnosed before coma occurs if recovery is to be achieved It is true that some cases have a very acute onset but these are invariably fatal

On occasion stupor and coma may be associated with acute encephalomyelitis such as occurs both as an illness in itself or associated with other diseases particularly those of a virus nature These comatose patients are always gravely ill and mortality is high but confirmation of the diagnosis in this acute phase which always occurs at the beginning of the illness is often pos

sible by lumbar puncture and is worthwhile as effective treatment is sometimes possible All such patients should be treated in hospital and it is in fact true to say that all cases of cerebro spinal infection seen in coma should be transferred to hospital with the least possible delay

This discussion of the local or cerebral causes of coma may conveniently be closed with a brief reference to coma associated with head injury This has already been partly covered as intra cerebral and subarachnoid haemorrhage occur with as well as without trauma When the history of a blow on the head is obtained as it usually is diagnosis is simple but nevertheless a careful examination of the head is advisable so that the situation and nature of any fracture may be determined This can be of value not only in relation to the immediate question of disposal and transport but also in the case of a summons to give evidence should legal proceedings follow the injury When no history is obtainable the examination is all the more essential It must of course be cautious as careless handling may initiate or increase intracranial haemorrhage The edge of the fractured bone may be felt and traced so as to demonstrate its lack of resemblance to the other linear depressions or ridges normally found on the skull Another feature suggestive of fractured skull is subconjunctival haemorrhage with local oedema

Profound unconsciousness following head injury is a serious prognostic sign as less gravely injured patients tend to be restless but nevertheless all must be transferred at once to hospital as no case is hopeless In some cases of extradural or subdural *bleeding however coma is a delayed event occurring* some days (though more commonly within forty eight hours) after the injury These patients are sometimes overlooked in the Casualty Department of a busy general hospital and can be missed by anyone The diagnosis should normally present no great difficulty if this possibility is borne in mind Following a head injury even if this appears to have been slight the individual should be put to bed and observed closely for three days This is of particular importance if he has been temporarily dazed or unconscious His behaviour should be noted as it is liable to be slightly abnormal in one way or another A tendency to drowsiness slowing of the pulse and variations in respiratory rate are signs strongly

indicating haemorrhage and should lead to admission to hospital or to a request for the advice of a neuro surgeon Squints and pupillary changes are further indications for urgent action but these are more likely to be absent Papilloedema valuable *as it may be at a later stage is not likely to be an early* sign time should not be wasted waiting for it and the same consideration applies to other neurological signs One further point may be made before leaving the subject In some cases unconsciousness is immediate but may be due either to drunken ness or concussion If it is decided to keep such a patient at home regular careful observation is necessary as in a few hours the patient may slip into deeper unconsciousness due to extradural or subdural haemorrhage and precious time may be lost

GENERAL CAUSES OF COMA

Pride of place must be given here to poisoning treated in this chapter in the general context of coma and from the viewpoint of the general physician The reader will find a more frankly forensic treatment of the same subject in Chapter XX In the Western hemisphere poison is by far the most frequent of the general as opposed to the cerebral causes of coma Analysis of 412 cases of poisoning admitted to a large provincial teaching hospital between 1952 and 1956 revealed 140 examples due to barbiturates and twenty four to other soporific drugs with similar effects Seventy seven cases were due to coal gas and carbon monoxide poisoning fifty nine to aspirin seventeen to corrosives seven to morphine and six to alcohol Miscellaneous poisonous agents accounted for the remaining eighty two cases Not all of these patients were admitted in coma but rather more than half were either stuporose or comatose

Suicide is the usual reason but accident is not uncommon especially among children No great amount of space can be given to this subject here but in all cases urgent admission to hospital is the wisest course

Gastric lavage when practicable should be attempted when ever delay in hospital admission is inevitable because of distance or other reasons but in a comatose child skilled assistance is usually necessary if more harm than good is not to be done and

certainly in the early stages induced vomiting is preferable Once other causes of coma have been excluded so far as is possible the most fruitful diagnostic line is an enquiry into the possibilities of access by the child to the stocks of poison in the household (and there are few households nowadays without such stocks) and a search for actual evidence of such access Information of this kind can be of the greatest value to those responsible for the treatment of the child in hospital

Accidental poisoning also occurs in adults of course now that the multiplicity of therapeutic tablets and capsules is outrunning the number of colours in the rainbow while the leaking gas pipe and faulty water heater are still with us A consideration of the numerous toxic possibilities from household cleansers to tranquillizers at once reveals the impracticability of an exhaustive account of the subject in the space available Only the more popular poisons already mentioned will be dealt with in their clinical aspects brief reference to others being made in passing where possible

The barbiturates in their various forms and combinations come first in order and can be treated together for our present purpose It is true that different barbiturates have different rates of absorption and duration of action and so may produce somewhat differing clinical pictures but this tendency is unimportant in the early stages of coma provided one is aware of it Anyway suicides often seem to take a variety of tablets some of which may themselves be complex and it must always be assumed in treatment that this may have occurred in the absence of evidence to the contrary This is not to say that knowledge which may help to identify remnants observed in gastric residues or chests of drawers is of no importance

The general course of events in barbiturate poisoning is drowsiness passing into stupor and coma but a stage of excitement restlessness or even delirium may precede or intervene Occasionally this dubious phase may be dangerous in that it may mislead the doctor and induce him to give sedative (*e g* barbiturate) remedies thus aggravating the condition The writer has once or twice known this to occur The barbiturates are depressants of cerebral mid brain and medullary activity and evidence of this depression is readily observed on examination After the

initial stage of excitement, all reflexes become depressed and then finally lost The pupils may dilate initially but they soon contract, to dilate again later In the early phase however their size tends to fluctuate The blood pressure falls steadily and the pulse is rapid Cyanosis is usually a late sign though secretion may collect at an early stage in the larynx causing some respiratory obstruction Sensory depression may best be judged by conjunctival stimulation and by the reaction to pin prick and muscle compression Loss of pain sensitivity may occur at a fairly early stage and depression of deep reflexes may be noted before they disappear Complete absence of deep reflexes is a sign of severe intoxication and of grave prognostic import if it is prolonged Many patients recover from this advanced stage with active treatment but its presence is a warning that time is short The superficial reflexes are of little value in this resp ct and the writer has not often seen extensor plantar responses

Death at this stage of the loss of deep reflexes (or even earlier) is due as often to circulatory shock as to respiratory or cardiac failure and the need for warmth is urgent in all cases of barbiturate coma Many of these patients are found lying out of doors or unclothed and it must be remembered that the maintenance of the body temperature is one of the earliest of the vital activities to be affected by the drug The spectacle sometimes seen in hospital of a comatose patient lying half exposed whilst earnest doctors carry out detailed examinations and engage in eager discussion illustrates how scientific inquiry may outrun common sense Another elementary danger is pharyngeal weakness which may allow the tongue to fall back into the throat

The position is however that barbiturate poisoning frequently does not present as coma of unknown origin Information is usually available from one source or another which indicates barbiturate overdosage and the doctor is in fact sometimes in danger of being misled by such evidence to overlook some other major causal factor in the patient s unconsciousness

A careful examination is therefore necessary and this should take account not only of the actual diagnosis of barbiturate poisoning but particularly of the stage and severity of the condition which may provide urgent therapeutic indications

The gas oven has been displaced from its once undisputed position as a cause of toxic coma but it still comes second in popularity and must be briefly considered The circumstances of the discovery of the patient inevitably give a clue as to the cause and diagnosis is thus not often in doubt Little need be said concerning the classical appearance but the feeble pulse heavy breathing, gross cyanosis and fixed contracted pupils with coldness of the body deserve mention In pure' carbon mon oxide poisoning e g in a closed garage the pupils are however apt to be dilated Oxygen should be administered during the journey to hospital which should be arranged as an emergency Spectroscopy is simple and confirms the diagnosis in any case of serious doubt

Poisoning due to aspirin is always suicidal in intent if one may judge by hospital records which reveal it as a popular but rather futile method If coma is achieved success may crown the patient s efforts but this is rare Of fifty nine patients admitted suffering from overdosage with aspirin all of them suicidal only five achieved coma two of whom died within a few hours of admission Furthermore beside one of these successful suicides lay an empty gin bottle Of the remainder many of whom had taken over 200 tablets only three were even drowsy The others were seen to be sweating but otherwise not affected There was only one instance of haematemesis in the series a point of interest in view of current opinion on the dangers of aspirin therapy In the patients with coma sweating was invariable as were the deep sighing respiration of acidosis and dilated pupils Reflexes were generally depressed but in one of the more severe cases extensor plantar responses were recorded The conclusion appears inescapable that aspirin is an ineffective method of committing suicide but that if the intention is serious a bottle of gin is a useful adjuvant

The Gerhardt test is a valuable aid to diagnosis It is true that the colour produced by the addition of a solution of ferric chloride to the urine resembles that caused by ketones but there is a definite purple tinge which should enable it to be dis tinguished from the Bordeaux red of diacetic acid

Corrosives are now relatively unpopular and commonly betray themselves by signs of burning of lips or mouth not to speak of the usual propinquity of the bottle

In morphine poisoning the signs and appearances resemble those of barbiturate poisoning except that the respiration is shallow and the pupils are fixed and severely contracted The condition is rare today but diagnosis is nevertheless most important since morphine antagonists are now avaliable Any patient in coma with pin point pupils and depressed or absent reflexes should be suspected of morphine poisoning and if *reasonable confirmation of such suspicion is found the adminis* tration of Nalorphine or admission to a hospital where such treatment is available is urgently necessary

Alcoholic intoxication is perhaps the most well known cause of coma but is probably less important today than formerly As all that is required in such cases is the provision of warmth and care and as the diagnosis is usually easily made little need be said from this point of view The flushed face dilated pupils heavy breathing and aromatic exhalation are readily recognized and the history is usually available It is most unlikely in fact, that the diagnosis will be overlooked The danger is rather that coma due to some other cause may be thought to be due to drunkenness perhaps because the breath may reek of alcohol *Furthermore the taking of alcohol may in fact precipitate coma* due to such causes as epilepsy or diabetes

THE ENDOCRINE COMAS

This is a large subject although it accounts for a relatively small proportion of comatose patients met with in practice The vast majority of such cases occur in diabetic subjects although nowadays insulin coma is much commoner than diabetic coma These cases must therefore occupy pride of place

Diabetic coma proper should not be difficult to recognize The deep sighing breathing (which is also rapid) the general flaccidity of all muscles with depression of deep reflexes and the soft eyeballs (though once the writer was almost led astray by a man with a glass eye!) are most suggestive—even if no history is available In fact of course this is a rare eventuality All these

findings are in marked contrast to the restlessness muscular irritability and exaggerated reflexes with extensor plantar responses so characteristic of hypoglycaemic coma Extensor responses are never found in diabetic coma

The initial diagnosis should be clinical as it occasionally happens (though infrequently) that both sugar and ketone bodies may be present in the urine in hypoglycaemia as well as in comatose non diabetic patients On the other hand examination of the urine is absolutely essential Every effort must be made to procure a specimen and if the bladder is empty a bottle bedpan or other suitable receptacle should be maintained in position until the objective is achieved However it must be stressed that clinical examination comes first in importance as in time The writer was once informed by his house physician that three cases of diabetic coma had been admitted within two hours He found that three comatose patients had been admitted and that the urine of all three undoubtedly contained an abundance of sugar and acetone In two however clinical examination revealed fever and cerebral signs spinal puncture in these patients revealed the final tally as one case of diabetic coma one of meningococcal meningitis and one of subarachnoid haemorrhage

The above description may have given the erroneous impression that the history or indeed any information gathered from relatives or friends is of little importance The clinical findings have been given first because it may well be necessary to make the diagnosis in the absence of any other reliable information But every detail bearing on the past history and more particularly on the course of events leading up to the onset of coma must be obtained as far as possible Most cases occur in known diabetics and the early symptoms prior to loss of consciousness in such cases are usually lethargy vomiting and deep sighing respiration while there may be a story of some acute illness But whether or not this is so once the diagnosis of diabetic coma is established a search for evidence of any acute infection is urgently needed So often is such evidence to be found and so important is its active treatment that no diagnosis of diabetic coma can be regarded as complete unless and until the presence or absence of a complicating illness has been clearly determined

The coma of hypoglycaemia develops almost always in known diabetics who are either careless injudicious or badly advised The history is of agitation restlessness hallucinations and perhaps convulsions It is important especially because cases are occasionally met with in non diabetic subjects Pancreatic tumours or overgrowths of the islet tissue may cause recurrent attacks which tend to be self limited and a history of these may be obtained They are sometimes mistaken for epilepsy but if the history is taken carefully this should be avoided Hypoglycaemia occurring in an epileptic person may provoke a fit and this does present a problem in diagnosis in a non diabetic But such an occurrence is rare

There are several non diabetic causes of coma with hypoglycaemia of endocrine origin which must be briefly considered These are uncommon but liable to present unexpectedly The most important are the adrenal failures and Addison s disease stands first In the established case the diagnosis is obvious but in the undiagnosed case and particularly that of the patient with little or no pigmentation difficulty arises Such a patient may be operated on or may sustain an acute injury or an acute illness and may pass into a coma for which no obvious cause appears In any such case when other causes are excluded (*e g* diabetes renal disease) an injection of cortisone may be life saving

The acute adrenal failure sometimes associated with meningococcal septicaemia is a real medical emergency and must be briefly mentioned despite its considerable rarity The patients often children are generally examples of severe or fulminating meningococcal septicaemia and show the petechiae associated with this affection In any child or young adult who is taken acutely and seriously ill with high fever and who collapses and becomes comatose at any early stage of the illness this possibility should come to mind petechiae should be searched for Meningitis will probably not be present. Massive doses of penicillin are of course required but intravenous hydrocortisone is also needed Prompt treatment is life saving hence the importance of the diagnosis The ordinary signs of hypoglycaemia are apt to be masked in these cases but the plantar responses may be extensor There may be vomiting for a short while prior to the onset of coma and this may be confusing but the gross

asthenia and hypotension cannot be accounted for easily by the moderate amount of sickness

A similar condition with or without injury may develop in a person with pituitary deficiency but here the scarcity or absence of axillary and pubic hair is of diagnostic importance

This brief review may be concluded by saying that any non diabetic person who passes into coma with or without obvious evidence of Addison s disease who has gross hypo tension and in whom no evident cause is to be found is likely to be suffering from acute adrenal failure This is more particu larly so if the onset of coma has followed an injury operation or acute illness

The thyroid comas need little more than a mention Coma occurs both in myxoedema and in thyrotoxicosis but in both it is rare Furthermore it is seen only in very advanced cases where the diagnosis is obvious and the prognosis very serious The sudden respiratory failure which may occasionally occur in myas thenic persons may lead to coma and is an urgent indication for hospital treatment but it is in fact most often encountered in patients already in hospital and is usually easily recognizable

BIOCHEMICAL COMA

This consists mainly of conditions covered by the general term uraemia The first and most important member of this group is of course renal failure This may have a sudden onset but the preceding symptomatology usually reveals its nature The ingravescent stage of chronic uraemia is so well known that it need barely be mentioned The anorexia the nausea and sickness with a bitter taste of urea in the mouth the minor twitchings and cramp and the high urine output are liable at any time to be terminated by coma more particularly in the event of an incidental acute infection The earthy pallor the slight puffiness of the eyes the hissing deep breathing the muscular irritability and the extensor plantar responses are all characteristic Con vulsions may occur Examination of the urine reveals albumin and casts though not always a large amount of albumin

The muscle irritability is tetanic in nature and may yield a positive Chvostek s sign but short of this it is readily demon

strated by the gentle application of the patellar hammer to the forearm when each individual muscle bundle responds in its turn by a twitch These finger twitches may of course be present without stimulation

In acute nephritis convulsions followed by unconsciousness may occur at the onset but this is due to acute hypertension rather than to uraemia The younger age of the patient and the short history are distinguishing features and the prompt recovery often seen following venesection or lumbar puncture is not a feature of renal failure of biochemical type

Next are the non renal acidoses whether respiratory or intestinal in origin and the alkaloses The acidoses are perhaps the more important Severe and persistent *diarrhoea* may lead to *acidosis* as indeed will any condition causing *dehydration* Coma may occur if the condition is sufficiently severe and acute or if renal function is impaired Respiratory acidosis is more important because it is not widely known difficult to recognize and yet not uncommon It has already been mentioned but must be briefly referred to again The chronic bronchitic with *emphysema* depends for his life on the already inadequate ventilation of an inadequate respiratory surface area Any acute bronchial infection may suddenly reduce this below his minimum requirement and carbon dioxide accumulates as oxygen saturation falls Thus acidosis develops and the respiratory centre becomes refractory to the normal stimulus Carbon dioxide narcosis develops and coma supervenes In such a case coma may also follow the administration of oxygen for reasons which cannot be discussed here The danger of giving morphine has already been mentioned Cyanosis twitching and even convulsions occur Clinical diagnosis is not difficult It is necessary only to be aware of the condition Urgent hospital admission is indicated

Any study of biochemical coma would be incomplete without mention of acute or chronic hepatic failure Cholaemia or acute liver failure is the mode of death in many infections involving the liver *e g* infective hepatitis Weil s disease and amoebiasis Sometimes it may be induced by an illness or operation in a starved individual but this is now rare The condition usually develops during an illness for which the patient is already under treatment and the diagnosis is simple provided the condition

is kept in mind Jaundice is usual but not invariable at the onset Various cranial nerve palsies muscular twitchings generalized convulsions and extensor plantar responses may all be manifest against a background of noisy delirium

In contrast are the patients with chronic hepatic fibrosis and extensive extra and intra hepatic portal systemic venous anasto moses Without apparent evidence of the underlying cause these patients may develop a relapsing stupor with preceding person ality changes and flapping tremor of the hands and fingers as the only indication of auto intoxication with nitrogenous breakdown substances by passing the liver cells As in acute hepatic failure so in most of these cases a careful history will reveal evidence of previous liver disease Clinical examina tion will demonstrate the flap foetor and exaggerated reflexes with extensor plantar responses in the comatose case which have come in recent years to be recognized as the signs of portal systemic encephalopathy Early recognition is important since the immediate prognosis is good if protein foodstuffs are withheld and the patient is given a broad spectrum antibiotic such as Neomycin

Certain of the signs of cholaemia are due to vitamin deficiency and it is convenient to mention here coma due to deficient vitamin intake This may still be occasionally seen and not only in the comatose stage of delirium tremens Occasionally in elderly ladies in straitened circumstances and poor health coma of uncertain origin occurs If careful study fails to reveal a cause intensive daily treatment with thiamine and nicotinic acid is well worth a trial though there may be no response for ten days or so

The coma of haemorrhage and anaemia may sometimes mislead if it is not borne in mind Cases of pernicious anaemia or melaena are sometimes admitted in coma which responds miraculously to treatment by vitamin B_{12} in the one case and to cautious transfusion in the other Rectal examination in a comatose person may reveal gastro intestinal haemorrhage as the cause by the presence of tarry melaena on the examining finger

Finally a cause of coma worth mentioning is that due to circu latory failure This is really in effect anoxia and the two examples

which come to mind are coronary thrombosis and the Stokes Adams disorder

EPILEPSY AND HYSTERIA

Epilepsy and hysteria are conveniently considered together Such consideration need not be prolonged

The condition of status epilepticus is readily recognized though it has to be borne in mind that an intracranial tumour may be present and that the condition is not necessarily a manifestation of idiopathic epilepsy But a condition of unconsciousness may succeed this or may replace it in an epileptic person The stertorous breathing which is noted in status epilepticus may persist for a while but it may not be present at all or may cease We then have an unconscious person moderately cyanosed who is breathing deeply and rather noisily and who may or may not show occasional twitching of the limbs The reflexes tend to be depressed or absent though the plantar responses may be extensor If as is usually the case this phase is of short duration diagnosis is neither difficult nor urgent but in the unusual case of prolonged unconsciousness doubt and difficulty may arise if the history has not been obtained or is unobtainable The exclusion of other possible causes of coma more particularly of the intracranial causes and insulin overdosage is clearly a first necessity

The hysteric is liable to be more of a problem Although hysteria is often very readily recognized the diagnosis is one always fraught with danger The hysterical patient may for example so accurately mimic a deep and dangerous coma and one moreover for which no cause can be found that considerable anxiety may be caused If blepharospasm hyperpnoea and the hysterical moan are noted little difficulty in recognition may be experienced But when immobility and insensibility are profound and the reflexes obtained with difficulty diagnostic deadlock is close at hand and the electroencephalogram may have to be invoked Fortunately the condition is not dangerous and the important aspect of the case is really the exclusion of other and more serious states The writer has encountered several such patients and found they were apt to be boastful afterwards and to retail remarks overheard by them during their coma

Hysterical coma is occasionally the forerunner of disseminated sclerosis and a watchful attitude is always advisable in the presence of such a history

THE COMATOSE PATIENT

The first essential in diagnostic method is the collection of all information which can be gleaned from relatives bystanders or neighbours regarding the immediate pre comatose period and the mode of onset of unconsciousness Was this abrupt and dramatic or was it characterized by gradually increasing drowsiness restlessness delirium or convulsions? Was there any accident or injury and was headache with or without vomiting a prominent feature? The question regarding injury is most important for an epileptic fit or a syncopal attack may lead to subdural haemorrhage A helpful history is obtained in most cases and the more energetic the enquiry the more likely is it to materialize The details so obtained are of value not only to the practitioner first called to the patient but to the hospital doctors who may be asked to care for him later they should be passed on with the patient if hospital admission is arranged

The more remote history must also be sought It will probably be known to somebody if the patient is a diabetic and if so whether he is taking insulin and particular enquiry should ascertain if he has met with any recent accident or illness and if possible the time and amount of his last insulin injection An epileptic is also usually known as such while a history of depression or of previous suicidal attempts on the part of the patient or relatives is often available for the asking in cases of coma of doubtful or suspicious origin

Clinical examination must of course be thorough in every case but it is considerably aided and guided by such information

The apparent age of the patient is to be noted A young person is unlikely to be apoplectic and embolism or endocrine coma is rather infrequent in the elderly In all cases the patient s colour and the character of the breathing will no doubt first be observed on general inspection The pallor of poisoning shock or haemorrhage the flushed appearance of apoplexy and epilepsy and the cyanosis of carbon monoxide poisoning and carbon dioxide

narcosis are characteristic Pigmentation of face and buccal mucous membranes points to Addison s disease while petechiae may indicate meningococcal septicaemia with adrenal failure Jaundice argues for hepatic coma though of course its absence does not exclude this

The deep sighing breathing of diabetic coma and uraemia or the stertor of the cerebro vascular accident or epileptic state contrast with the shallow breathing of most forms of poisoning and the exaggerated hyperpnoea of the hysteric Irregular and intermittent breathing such as the Cheyne Stokes phenomenon usually indicates intracranial coma It also occurs however in respiratory disorders associated with anoxia such as respiratory acidosis and in biochemical disorders *e g* uraemia It is very easily induced in elderly people

Palpation of the pulse is often of little value in the diagnosis of coma (it tends to be of low tension except in apoplexy) but it is helpful to know that it can be felt and in cases of coma due to heart block the abnormally slow pulse may yield the diagnosis The presence of auricular fibrillation may help in the diagnosis of a cerebral embolus

The temperature may be of little help and in the case of a person found in an exposed situation it may be misleading But *a high temperature in an unconscious person always suggests* the presence of a general or intracranial infection and should initiate a search for other evidence pointing in these directions

Furthermore a deficiency in pubic and axillary hair may give a clue to pituitary coma unless the appearance of gross myx oedema indicates an alternative explanation

In the more detailed examination of the patient the nervous system must have pride of place Not only are the vast majority of comas intracranial in origin but nervous signs of importance are to be observed in coma of general origin The presence of local nervous signs such as lack of normal tone in a limb or limbs with changes in the reflexes or neck stiffness and muscle *rigidity is strong evidence of intracranial disease and* those cranial nerve defects which can readily be recognized in the unconscious patient (squints ocular deviations and facial para lyses) may provide further confirmation It may also be remarked at this point that failure to observe papilloedema may cause vain

regrets at a later date Some cerebral comas have no localizing signs however while in coma of extra cerebral origin ocular and pyramidal tract signs may occur Certain poisonings as well as some biochemical and nutritional comas are illustrations of this Uraemia and cholaemia are well known examples

The state of the pupils is of importance in most cases of coma Unilateral changes usually indicate intracranial damage but bilateral changes are commonly met with in other disorders Severely contracted pupils may be seen both in cerebral haemorrhage and in morphine poisoning but widely dilated pupils suggest either poisoning in a dangerously advanced stage or hysteria Variations in size of the pupils over a period are also of value in diagnosis and prognosis

General diminution of reflexes is common in most non cerebral comas particularly in diabetic coma and poisoning and complete loss of deep reflexes is a grave sign in both The reflexes may be exaggerated in insulin coma though not in hypoglycaemia due to causes other than hyperinsulinism

Examination of the heart may reveal the diagnosis in coma of embolic origin and the stethoscope is also valuable in the recognition of respiratory coma by the discovery of a valvular lesion or of gross ventilatory disturbances as the case may be Cases occur in which rectal examination discloses the presence of a melaena which may explain the occurrence of unconsciousness In most cases the urine must be examined as soon after the clinical examination as possible and if this need is accepted the specimen can practically always be obtained A haemoglobin estimation is sometimes of diagnostic value and finally lumbar puncture should not be undervalued as a diagnostic measure in any comatose patient in whom it is not contraindicated by evidence of severely raised intracranial pressure or suspicion of a posterior fossa lesion

CHAPTER XVII

PSYCHOSIS

By Desmond Curran

A GENERAL physician might well feel daunted if he were asked to attempt a short chapter on the early diagnosis of potentially serious illnesses met with in general medicine The task of the present writer is in many ways one of comparable difficulty and he hopes this may in some degree extenuate the inadequacy and superficial dogmatism of what follows

NEUROSIS AND PSYCHOSIS

Medical students often ask What do you mean by psychosis and how does it differ from a neurosis? or the more specific question Is this a case of neurosis or an early psychosis?

Common concepts on these topics can be summarised as follows

Mental disease without treatment becomes progressively more severe with the passage of time Early psychosis therefore implies a relatively mild condition of recent onset which if untreated may progress to insanity Secondly the psychoses are the major mental illnesses or insanities in which the total personality is involved in a serious and sweeping way The neuroses are the minor mental ailments or disorders not so serious in which the personality is only partially involved The neurotic is neither mad nor likely to become so Thirdly the psychotic lacks insight His judgement of reality is seriously distorted in some way He suffers for example from delusions or misinterpretations He lives in a world that is qualitatively different—his world is not our world Hence the popular story The neurotic builds castles in the air the psychotic not only builds castles in the air but inhabits them and the psychiatrist draws the rent Furthermore in the matter of causation genetic and constitutional or as yet unidentified physical factors are of major importance in the

psychotic as opposed to psychogenic factors in the neurotic illness Diagnostically schizophrenia and the manic depressive illness are psychoses hysteria anxiety states and obsessional illnesses are neuroses But the organic syndromes do not fit into the neurotic psychotic dichotomy and cause doubt and difficulty *e g* Would you call or when would you call a senile illness psychotic? Finally from a therapeutic viewpoint psychotherapy is held to be the major weapon in neuroses some physical method of treatment such as electroconvulsion insulin or chlorpromazine (Largactil) in the psychoses

THE CRITERIA FOR PSYCHOSIS

According to the views outlined above the three main criteria for psychosis are *severity* in the sense of being certifiably insane *insight* as judged by the presence or absence of delusions hallucinations or misinterpretations and the medical *diagnostic category* with special reference to schizophrenia and the manic depressive illnesses

Application of these three criteria does not however always lead to the same answer Whilst it is true that some patients especially certain schizophrenics may qualify as psychotic by all three criteria a very large number of patients only qualify as psychotic according to one For example many endogenous depressives (psychotic by the diagnostic criterion) are not certifiable and have excellent insight Again certain hysterics (neurotic by the diagnostic criterion) have quite properly been certified Conversely many paranoid schizophrenics with delusional ideas (psychotic by the criteria of diagnosis and insight) are not certifiable

Furthermore it is a serious mistake to equate psychosis with insanity or to suppose that patients classed as psychotic by the medical diagnostic criteria just mentioned will if untreated necessarily or even probably become certifiably insane To be certifiably insane is not a medical diagnosis Nor is insanity an entity It is a judgement concerning the need for compulsory care supervision and control sanctioned in certain circumstances by the law for the benefit of the patient and the protection of society

EARLY PSYCHOSIS

The term early psychosis is ambiguous It may mean psychosis either of recent origin or of mild type Gradual progression in severity with the passage of time is no more the rule in psychiatry than it is in other branches of medicine In fact as in other branches of medicine early illness in the sense of illness of *recent onset can be acute and recoverable and early illness* in the sense of being mild is sometimes of insidious origin and poor prognosis

The writer will confine his attention in this chapter to psychoses that are early in the sense of being mild and where the differential diagnosis from neurosis arises He will not fidget over definitions and will include organic mental syndromes amongst the psychoses

THE DIAGNOSIS OF NEUROSIS OR 'PERSONALITY PROBLEMS'

The diagnosis of neurosis tends to be a diagnosis by exclusion Thus having excluded physical disorder or disease as the cause of symptoms the doctor is inclined to lump all functional cases under the *genus* neurosis *species* anxiety state unless a gross disturbance of behaviour of psychotic intensity is shown

The term personality problem is in many ways preferable to either neurosis or anxiety state since it lays emphasis on what is often the most important thing namely the personality of the patient rather than the presenting symptoms Evidence that the patient is a neurotically predisposed or vulnerable individual liable to react to stress by the development of neurotic symptoms is usually demonstrable in the life history of cases correctly diagnosed as suffering from a neurosis

The three main psychiatric syndromes often wrongly diagnosed *as neurosis are the manic depressive group of illnesses at all ages* beyond adolescence schizophrenia in the younger age groups and organic syndromes especially in older patients such as those with arteriosclerotic and presenile dementias

Possible reasons for such mistakes are first the equation of psychosis with insanity As already pointed out psychosis is

psychotic as opposed to psychogenic factors in the neurotic illness Diagnostically schizophrenia and the manic depressive illness are psychoses hysteria anxiety states and obsessional illnesses are neuroses But the organic syndromes do not fit into the neurotic psychotic dichotomy and cause doubt and difficulty *e g* Would you call or when would you call a senile illness psychotic? Finally from a therapeutic viewpoint psychotherapy is held to be the major weapon in neuroses some physical method of treatment such as electroconvulsion insulin or chlorpromazine (Largactil) in the psychoses

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It should be more widely appreciated that only a small minority of such illnesses—in one series less than six per cent—ever show a manic or hypomanic phase and that the diagnosis of these illnesses very seldom implies a life sentence of frequently recurrent oscillations between the two phases

The hypomanic syndrome is consequently quite rare The main features are an unwonted push of activity talk planning extravagance aggression intolerance and argumentative quarrelsomeness that is sustained although not necessarily severe Without an independent history that permits of a comparison with the patient s pre morbid personality these patients may merely be regarded as difficult uncontrolled tiresome and psychopathic individuals or if secondary alcoholism is present the behaviour may be considered as solely due to this cause Usually it is the friends or relatives rather than the patient who seek help and advice over rows extravagance or failure in judgement Many of these patients can give a misleadingly normal impression in an interview To live with them is to know better Lack of insight is highly characteristic and makes treatment extremely difficult

The endogenous depressive syndrome is very much more frequent and important Under this heading will be included cases where this illness first appears during the involutional period of life It is important to be familiar with this syndrome because it is very common amongst adults of all ages and is often wrongly diagnosed or missed while the results of treatment are on the whole very good

The presenting complaints are often of a physical kind The patient feels rotten and looks ill Physical disease is therefore suspected Subjectively anxiety tension and fears are an integral part of many underlying depressions combined with the somatic accompaniments of these emotions This often leads to the diagnosis of neurosis Or secondary reactions of anxious hypochondriacal or hysterical type may develop to the underlying depression just as they can develop to physical disease *These features may then dominate the picture The self* absorption and importunacy so common in depressions often seem typically neurotic

apt quite wrongly to be equated with insanity with a consequent reluctance to diagnose a psychosis unless a severe degree of disturbance is obvious Secondly somatic complaints in psychosis *give rise to difficulty The presenting complaints in all psychiatric* conditions are very commonly of a physical type That somatic manifestations or complaints—headaches palpitations aches and pains fatigability—are very common in neuroses is universally recognized but it is not so widely appreciated that exactly the same somatic complaints also commonly occur in the early psychoses under consideration here Spurious psychogenesis is also a common pitfall It has been pointed out that constitutional or physical aetiological factors are regarded as of major importance in the psychotic as opposed to psychogenic factors in the neurotic Whilst this is true by and large the basic aetiological principle in psychiatry is that of multiple causation and the issue is one of emphasis and not antithesis But we are all tempted to look *for the cause and when this seems to be psychogenic we* may be further tempted to regard the condition seen as essentially psychogenic and hence neurotic Again we may be reluctant to make a diagnosis of what we may regard as tantamount to the diagnosis of incipient insanity when a natural reaction to stress seems both more plausible and more palatable

Finally it may not be known or fully appreciated that it is unusual for an individual with a good personality and work record to develop neurotic symptoms except as the result of *exceptional and obvious stress or that it is most unusual for* any patient to develop neurotic symptoms for the first time after the age of forty except in the above circumstances

MANIC DEPRESSIVE PSYCHOSIS

The essential feature in both phases is the sustained disturbance of well being that is shown persisting for weeks or months

It is important to emphasize well being rather than mood disturbance in the direction of either depression (melancholia) *or elation (mania) Paradoxical as it may seem many depressive* patients do not complain primarily of any depression of mood and they may even deny it while equally many manics or hypomanics are not happy or elated but over active and aggressive Excitement is therefore a better descriptive term than elation

poverty ideas associated with agitation rather than slowing up or retardation He can t settle to anything at all

It is highly characteristic for all symptoms of depression at all ages to be *worse in the mornings* and to improve as the day goes on and in the evenings (diurnal variation)

When the picture is one of hopeless gloom and doom the diagnosis is not difficult and the possible suicidal risk obvious All that is seen in many cases however are the symptoms of loss previously outlined associated with inertia and negativistic indecision and querulous hypochondriasis none of the symptoms necessarily being severe

Serious errors of judgement can result from a depression as well as from hypomania Jobs may be resigned because of a sense of failure and inadequacy or houses sold because of un justified ideas of poverty or pessimism concerning future prospects

SCHIZOPHRENIA

The older term dementia praecox is misleading since the onset need not be early nor the prognosis necessarily bad

Schizophrenia includes a group of grave common and usually progressive mental illnesses that occupy nearly one in five of all hospital beds These mental illnesses are characterized by a tendency of the patient to lose effective contact with ordinary life and to withdraw into an inner world dominated by phantasy and delusion In severe cases the end result is a condition of profound mental deterioration with the patient unoccupied in accessible and apathetic but without that clouding of conscious ness or true intellectual impairment which is seen in organic syndromes such as senile dementia During the early stages the clinical picture can vary The onset may be sudden and the symptoms florid In general when this is so the outlook is better We are concerned here however with cases of a milder type although not necessarily with a better prognosis which may simulate depression neurosis or personality problem

The presenting complaints may be what the patients them selves complain of or what they do or do not do that is com plained of by others than the patient who often take the initiative in seeking medical advice

In order to assess the presenting complaints the associated symptoms to be looked for can be considered under two headings physical and mental

The sustained depression in vital activity is more essential for the diagnosis than the actual depression in mood Thus a persistent lowering of energy with a consequent lowered drive and increased fatigability is shown Activity is thus reduced In addition not all patients show all but all show some and many most of the following more specific biological con comitants of an endogenous depression—loss of appetite weight sleep and sex desire and constipation The character of the sleep disturbance is not so much difficulty in getting off to sleep as in *broken sleep with early waking*

The mental symptoms run parallel Thus the reduction in physical activity is accompanied in the mental sphere by a feeling of being unable to cope as well as usual coupled with a disinclination to do as much as before The patients are often described as slowed up The keynote is *loss*—loss of interest and pleasure loss of confidence and decisiveness loss of energy and drive With the loss of interest the patient cannot experience enjoyment The ordinary pleasures of the outer world leave him cold and he may also be distressingly aware of his loss of feeling for his loved ones so that he feels emotionally dead (affective loss) The loss of interest in the outside world contri butes to self centred self absorption and with the loss of decisiveness comes a tendency to procrastinate

The combination of inability to cope indecision and self absorption leads to a sense of inadequacy and thus to self depreciation and *self reproach* Self reproach—often described by relatives as an inferiority complex or guilt complex —is a classical symptom but one not invariably shown When it is experienced patients may come to believe that others must feel as they do about themselves and that they are being looked at and talked about in a critical and disparaging way (ideas of reference) They may feel this is justified or they may resent it

In the older age groups hypochondriacal concern over their bodily functions or financial position becomes increasingly prominent and may progress to classical bowel blocking or

apparent reason and when quite sober had suddenly heaved a brick into the window of a police station was equally stunned by his action and by his subsequent attitude— What a thing to do! And he wouldn t tell me why he did it and didn t seem to mind And he has always been such a quiet well behaved boy And I know he hadn t any friends to put him up to it This bizarre incongruous quality can also be seen when patients who spend most of their time in what is apparently brooding silence or depression are observed to smile or even giggle to themselves for no apparent reason

The volitional disturbances are seen in the drop in efficiency and drive and the lack of energy and determination To quote another relative He seems to have lost his initiative and go You can t get him up in the morning and he won t shave properly He seems to have lost his pride It is obvious how closely the volitional and emotional disturbances are intertwined

Delusions and misinterpretations are common The essence of what are called primary delusions is that they spring up suddenly and fully armed or developed with an unshakeable quality of conviction Chance remarks or chance happenings possess an intense personal significance or meaning The traffic lights turn green or red in succession this can only mean you are specially singled out for favour or persecution It is not mere chance that when you enter a room everybody starts to laugh or to smile The facts are indubitable that they mean something specially directed at you is certain but it may not be quite clear what the meaning is Clearly you are being tested in some way—but why and for what purpose? Secondary delusional elaborations of a more crystallized type may occur Your husband is unfaithful that piece of paper he tore up proves it he would not have looked so guilty if it had not been a letter from his mistress

It is the quality of the evidence adduced by the patients to justify their suspicions that is significant Your husband may in fact be unfaithful your employer in fact may want to get rid of you and your neighbours do perhaps dislike and shun you But this cannot be deduced with certainty from a torn up piece of paper or because your employer looks the other way and coughs when he sees you or from the observation that when

A gradual change is observed in the direction of increased in effectiveness oddness or eccentricity often accompanied by non specific symptoms suggestive of physical ill health or neurosis The patient does less and less and becomes increasingly self absorbed hypochondriacal and seclusive The outward impression may be that of a difficult idle solitary stubborn youth or damsel who is egocentric and cold but at the same time hypersensitive touchy and unpredictable Amongst th more intelligent an interest in esoteric religion or philosophy is common

The falling off in efficiency energy and powers of concentration especially when hypochondriacal self concern is present may be attributed either to physical causes (to which a pale weedy appearance often lends colour) or to psychogenic stress (such as love affairs gone wrong or examination failures) The attitude of parents—often themselves odd and difficult people—may be one of defensive perplexity *e g* It can t be serious can it? or I give him up I have never been able to understand him I think he justs needs to get down to a job of work

The associated symptoms in schizophrenia are classically described as bizarre This indefinable quality of being un accountable and odd and somehow frightening is highly characteristic The thought disorder one of the cardinal symptoms is easier to recognize than to describe but in answer to enquiry relatives may report that it has become increasingly difficult to follow what the patient means and that answers to questions have seemed off beam and beside the point or woolly Spontaneous remarks may take on an alarming out of the blue quality As one relative put it He says things at times I cannot understand and which seem to have no connection with what we have been talking about It really makes me wonder if I am losing my reason—but he is so clever and I am not I expect that s it

The emotional reactions tend to become inadequate or in appropriate or both Thus neither explanation nor excuse may be *given for idleness failure at work or anti social acts* This is in contrast with the behaviour of psychopaths who usually try to trump up some excuse or invent plausible lies to explain their failings Thus the mother of a patient who for no

When these are present organic causes when not otherwise obvious should be sought

The term organic deterioration is preferable to organic dementia until the cause has been determined Organic dementia should strictly speaking be confined to an *irreversible* decline of mental function produced by organic disease of the brain *e g* senile dementia But exactly similar mental symptoms can be seen in potentially recoverable cases *e g* those with meningiomata myxoedema or chronic intoxication by drugs

The basic symptoms in organic mental syndromes are evidence of deterioration of the personality predominantly in the intellectual sphere but also in the character

They fall under three main headings Intellectual deterioration is shown in slowness and difficulty in grasp impairment of memory especially for recent events and by failure of efficiency at work Organic lability of mood implies emotional incontinence the emotional flood gates are easily opened by stress These emotional reactions are comparable in less vivid form and slower tempo with those of the drunken man whose mood rapidly changes from bonhomie to aggression or to lachrymosity Organic mood changes often have a certain childish or petulant quality are typically highly reactive to circumstances and are transient rather than sustained Changes in character are shown by unexpected errors of judgement, deterioration in thoughtfulness personal appearance and manners and by a surprising indulgence in anti social acts that are out of keeping with the previous personality Such personality changes can occur without much intellectual impairment or vice versa

In some instances however the above general (and to that extent impersonal) organic mental symptoms may be masked by secondary reactions of anxious depressive hysterical or hypochondriacal type largely dependent upon the individual make up of the patient and his life situation Such secondary syndromes may indeed determine the symptoms with which a case of mental deterioration initially presents to the doctor and in such circumstances the underlying organic condition may well be overlooked

Organic syndromes are often due to degenerative changes and hence the main incidence is in the later decades of life The usual age of onset for *hypertensive disease* is between forty five

ever you leave your house your neighbours all bolt into theirs or drive off in their cars with noisy ostentation and supercilious smiles

Passivity feelings are highly characteristic Your thoughts can be read by others or are under some outside control Some form of hypnotism or telepathy is at work—thoughts are put in your mind This last experience shades into auditory hallucina tions Hearing voices in a setting of clear consciousness is usually due to schizophrenia

Paranoid schizophrenia and paraphrenia constitute rather a special form In many cases with paranoid delusional ideas and misinterpretations the personality is well preserved This means roughly that it is possible to meet such patients casually and notice nothing obviously the matter They are well kempt they are not in the least vague dreamy or withdrawn on the contrary they may be alert and quick and their emotional response (affect) not obviously flat or blunted or facile or silly The improvement with large doses of largactil can be striking

Schizophrenia is a very grave illness but the prognosis in any individual case is extremely difficult to predict An insidious onset is always ominous In round figures two out of three patients do badly and if followed up ten years later are in mental hospitals or are social invalids or have died

No organic pathology for schizophrenia has as yet been demon strated and its causation remains obscure The one definitely known fact in aetiology is that predisposition to schizophrenia increases rapidly with the nearness of blood relationship to a known case While environmental factors can play a part in precipitation no means of preventing the illness are known In general parents can be genuinely reassured that the illness is not due to the way their children were brought up

ORGANIC SYNDROMES

The organic mental syndromes comprise the organic deteriora tions and dementias in which physical factors are of predominant importance When this is so characteristic mental symptoms are shown These are known as organic mental symptoms

extremely well On the other hand, if the personality is well preserved a gross degree of dementia may not be found unless it is carefully sought Thus a dear old lady with many hypochondriacal complaints (which had led to exhaustive physical examinations including an unnecessary laparotomy) appeared neat in dress and quick enough to talk to but when asked the date would reply (rather testily) that she had not brought her Almanack with her In 1939 she believed that the year was 1906 several generations of medical students completely missed her dementia by omitting this quick and simple test

The mental symptoms in organic mental syndromes are in the main non specific They are more clearly referable to the intensity and duration of the morbid process than to the nature of this process Judged solely from the mental symptoms shown all that can usually be said is that they are of an organic mental type and although of course the most probable cause in a senile subject is brain decay the differential diagnosis of the physical factors at work depends upon a knowledge of general medicine supplemented by physical examination and special investigations such as serological tests for syphilis spinal fluid examination electro encephalography angiography and air studies

DIFFERENTIAL DIAGNOSIS

Many of the symptoms shown in psychiatric conditions seem normal enough when taken singly We have all personally experienced some degree of fatigue anxiety depression or difficulty in concentration Abnormality when it exists consists in the intensity and duration of such symptoms their combination together with other symptoms and the circumstances in which the symptoms arise

We have also seen that the presenting complaints in early psychosis are often non specific and of an apparently neurotic type The basic underlying symptoms of the psychosis in question may not be volunteered and therefore may not be found unless specifically sought

Some useful general questions to ask are does the breakdown come as a surprise? Is there a marked discrepancy between complaint and performance? Are the complaints persistent? Or

and fifty five for *arteriosclerosis* between fifty five and seventy *Senile dementia* rarely begins before the seventieth year But there is a group of cases known as the *presenile dementias* in which a slowly progressive dementia essentially similar to senile dementia starts in the fifties or even before Other causes of organic deterioration in the younger age groups include *cerebral tumours* and *cerebral syphilis* (the latter nowadays rare) and *chronic intoxications* by drugs and alcohol *Chronic uraemia* must also be mentioned

In many of these organic mental syndromes physical signs may be absent and the results of physical examination negative In others a careful physical examination may not have been carried out The breakdown or failure in efficiency may then be attributed to any secondary reaction shown For example the diagnosis of a reactive depression attributed to over work may be proferred and accepted when the depression is in fact reactive to failing efficiency due to organic mental causes aided and abetted by organic emotional lability

The clue to the correct answer must often be sought in the past history or biography bearing in mind first the trap of *spurious psychogenesis* and secondly *the relative rarity of a neurotic reaction developing for the first time in later life* except as a result of quite exceptional and obvious stress

Independent information from outside sources is usually essential in order to determine whether for example inefficiency at work or increased slowness and difficulty in grasp or memory impairment in fact preceded rather than followed the development of depression or anxious hypochondriasis

An additional reason for this emphasis on independent accounts is that the usual tests for orientation (knowing the date etc) and for memory retention and grasp (*e g* the retention of digits and other items the hundred minus seven test and so on) may be too crude In particular intelligent patients can often do well on tests of this kind even though their mental deterioration and lack of judgement may be obvious from their performance in life For example a regular Naval Officer after a severe head injury got into disciplinary difficulties that culminated in his attending Divisions quite imperturbably smoking a pipe Yet on tests of intellectual capacity he did

in the persistence of the complaints and the discrepancies between complaint and performance Prolonged observation may be necessary before a confident opinion can be expressed

The assessment of suicidal risk or threats is so difficult and complex that the opinion of a specialist may be considered desirable

Endogenous and Reactive Depressions

The term reactive when applied to depressions has led to confusion since it has been applied indiscriminately both to neurotic depressions in which a patient normally with a neurotic personality has reacted with depression to events he found stressful and to an endogenous depressive syndrome *precipitated* by stress

The differentiation is real and important neurotic depressions are not helped by electroconvulsive therapy (E C T) endogenous depressions (and it is immaterial whether they have been precipitated by stress or not) are The important contrast is the continuance of reactivity to environmental circumstances in the reactive or neurotic depression compared with the relative lack of reactivity to circumstances in the endogenous depressive syndromes

In reactive depressions a long history of neurotic symptoms can usually be obtained in the form of marked and rapid fluctuations in energy and interest a ready tendency to be moody and fed up and a general instability of mood and well being The more the symptoms vary in intensity from one moment to another the more capable the patient seems to be of snapping out of it when the telephone rings or there is talk of going to a party *the more likely it is that the condition is a personality* reaction or neurosis Again in the reactive depressions that are the direct result of special stress rather than expressions of more general personality dissatisfactions the symptoms usually follow upon the stress quickly and comprehensibly the patient is primarily concerned with the causal event or situation and an improvement in the external situation leads to a more rapid recovery For example the wife of a pilot learns that his plane is missing becomes depressed as a natural reaction to this news but rapidly recovers on hearing he is in fact safe and well The

are they highly reactive and variable? Can the patient appear perfectly normal in certain circumstances for considerable periods?

The question as to whether the breakdown comes as a surprise must be broken down and a distinction drawn between the fact of a breakdown coming as a surprise to those who know the patient and the symptoms shown or the type of breakdown Can the symptoms shown be construed as a normal response to stress given some knowledge of the patient and the stress or not?

If in the light of adequate information the fact of a breakdown occurring at all should come as a surprise the doctor should (after exclusion of obvious organic disease) suspect the possibility of either an organic syndrome especially when impairment of memory and grasp are prominent or a condition belonging to the manic depressive or schizophrenic groups This applies especially for example when an inexplicable loss of efficiency occurs in a previously effective person or when loss of confidence or hypochondriacal preoccupations develop in a person not previously subject to them

As regards the type of symptoms shown it is to be expected that somewhat unstable highly strung individuals should in certain circumstances develop anxiety emotionalism poor control or a variable degree of depression and sleep disturbance nor would we be unduly surprised at the development of such symptoms in apparently stable individuals as a consequence of severe and obvious stress But we as doctors should not expect intellectual impairment sustained expressions of guilt or other than the transient misinterpretations so often shown by touchy people When persistent intellectual impairment persistent guilt or persistent misinterpretations occur even after severe stress we should suspect the development of specific illness (or psychosis) rather than expressions merely of problems of attitude or personality (or neurosis)

But of course a psychosis can also develop in a neurotic unstable or vulnerable personality and also in dullards The symptoms of the underlying psychosis may then be coloured or obscured by the patient s personality or poor intelligence Even so it is the contrast with the previous state that must be sought

onset is therefore much more recent and abrupt and the response to E C T may be excellent

The secondary reactions of anxiety and depression which may occur in organic syndromes although they may appear quite severe at interview are more reactive to circumstances and do not have the sustained quality seen in the endogenous depressive syndrome

TREATMENT

As in other branches of medicine the practical importance of early diagnosis in psychiatry is closely related to the possibility of effective treatment The immediate disposal and treatment is usually based on the initial diagnostic formulation even though this may be admittedly provisional Cautious trial of a particular form of treatment may itself make an important contribution to accurate diagnosis

In general manic or hypomanic patients should be admitted to hospital to prevent foolish and extravagant behaviour to minimize aggravating stimulation and to substitute skilled and patient management for the efforts of their sorely tried families Out patient care on the other hand may be appropriate for depressive patients when social conditions permit and the risk of suicide is not too great Despite a contemporary swing of the pendulum against hospitalization of the schizophrenic almost all of the younger patients should be admitted for diagnosis assessment and treatment this is less necessary in older and more paranoid cases Admission of patients with organic dementia carries the risk especially in the older age groups that the family ranks may close with relief once the patient has left home and that the relatives may be reluctant to shoulder the burden again It is nevertheless often necessary for definitive diagnosis

Insulin coma is widely regarded as the treatment of choice for the younger schizophrenic patient with an illness of less than two years duration between thirty and sixty comas may be required and the patient must be in hospital for at least three months This treatment is often combined with E C T

It may be mentioned in passing that the type of treatment chosen must be left to the expert in charge of the case It is always a mistake to advocate strongly to relatives a special form

topics of preoccupation in an endogenous depression precipitated by stress are less sharply focussed and the endogenous depressive symptoms previously noted will also be evident

Fortunately the crucial contrast between the patient s state when ill and when well is often rendered easier in endogenous depressive syndromes by the fact that many of these patients are of good personality the last person one would have expected to have a nervous breakdown ' The change from a normal state of energy interest extraversion enjoyment confidence and decisiveness to one characterized by relative anergia loss of interest self absorption joylessness with lack of confidence and indecisiveness may then be immediately striking providing that the comparison is sought

Schizophrenia

It may be worth while stressing just one point again The presenting complaints are often apparently neurotic or merely suggestive of inadequacy Patients with quite severe distortions of content (delusions misinterpretations passivity feelings) may complain only of anxiety or depression which they regard as the natural consequence of the odd things going on The underlying schizophrenic symptoms may not be volunteered Unless given a chance to talk or specially questioned such a patient may keep his delusional ideas or misinterpretations to himself

Organic Syndromes

A common mistake in the aged is to diagnose a permanent organic dementia when in fact one is dealing with a potentially benign depression occurring in old age or benign mild confusion of exogenous origin (*e g* due to urinary infection)

The symptoms of a depression in the elderly or aged may simulate an organic dementia very closely owing to the slowness and difficulty in grasp that is often shown due in fact not to dementia but to depressive preoccupation and retardation The *duration of the illness* is often a helpful diagnostic point In senile psychoses the onset is slow and insidious with a history extending over many months or years In elderly depressions the history of change only dates back for weeks or months The

CHAPTER XVIII

MENTAL DEFICIENCY

By R S Illingworth

THERE are two main reasons for the importance of early diagnosis in mental deficiency Certain forms of mental deficiency are amenable to therapy only if treatment is promptly instituted Of these conditions the most important is *cretinism* The earlier thyroid deficiency is diagnosed the better are the chances that a normal level of intelligence will be achieved It must not be *assumed that all cretins will develop normal intelligence if* treated without delay not all will but the number will be greatly reduced if treatment is delayed

Another form of mental deficiency which should be diagnosed early is *phenylpyruvic oligophrenia* a metabolic condition in which phenylpyruvic acid is excreted in the urine and which is associated with mental deficiency It is now thought that if these cases are treated early by a special phenylalanine free diet the intellectual capacity can be at least considerably improved and that deterioration can be prevented Treatment should begin in infancy and certainly not later than the second birthday Most but not all of these children are fair haired There is often a family history of mental deficiency which may or may not have been recognized as being due to phenylpyruvia The diagnosis is readily made by acidifying urine with acetic acid and adding 5 per cent ferric chloride If phenylpyruvic acid is present in the urine a blue colour is immediately obtained The test can be applied readily to a freshly wet napkin It is now a routine test in all cases of mental defectiveness found in infancy

There are certain other metabolic conditions in which early diagnosis may be of value with regard to future intellectual capacity These include *galactosaemia* characterized particularly by severe neonatal jaundice with hepatomegaly and *idiopathic hypercalcaemia* diagnosed by the finding of a high serum calcium

of treatment that may not be available or may be considered unsuitable

Electroconvulsive therapy is usually required in endogenous depressive syndromes of any severity and the results are in general excellent Cure has been claimed in 70 per cent and great improvement in another 26 per cent In this connection especially it is clear that early and accurate diagnosis is imperative indeed many of the failures laid at the door of this form of treatment are due to its administration to wrongly diagnosed patients with neurotic depressions hysterical personalities or obsessional illnesses Heart failure and recent cardiac infarction are the main physical contra indications

Prefrontal leucotomy is a last resort in intractable states of tension or in patients with severe depression which fails to respond to E C T or which rapidly relapses

Amongst the drugs in common use the amphetamine group is generally disappointing in depressive illness tending further to increase tension and sleeplessness though 5 mg of Drinamyl in the morning may benefit some mild anergic depressions such as those which follow influenza Chlorpromazine (Largactil) useless in neuroses benefits some depressive patients awaiting E C T occasional hypomanics and many schizophrenics especially those of paranoid type As a sedative in old age also it has the advantage unlike the barbiturates of not adding further to mental confusion

Since the topic is early diagnosis the Editorial blue pencil has quite properly been employed to expunge any further discussion of treatment which of course consists of a mixture of psychological physical social and environmental measures in varying proportions according to the case

At five months he can go for an object and grasp it with his hands and at six months he passes an object from one hand to the other He begins to chew At seven months he can sit on the floor for a few seconds without support He begins to imitate actions and noises

At nine to ten months he can creep and pick up a small object the size of a currant between his forefinger and thumb By the age of a year he can say three words with meaning and in another month can walk without help He deliberately throws objects such as bricks one after the other on to the floor He has largely stopped slobbering and taking objects to his mouth though both these may continue for a few weeks after the first birthday in some normal babies By fifteen months he may ask for his pottie when he wants to empty the bladder and can pick up and put down a cup without spilling thus feeding himself From twelve months he has been helping to dress himself by holding out his arm for a coat and his foot for a shoe At twenty one to twenty four months he begins to put words together to form short sentences

These are the principal milestones and the average ages at which they are reached An understanding of these is essential for the early diagnosis of mental deficiency

THE BASIC PRINCIPLE OF DIAGNOSIS

The essential principle on which the early diagnosis of mental deficiency is based is the fact that the mentally defective child who is defective from birth or before birth is *backward in all fields of development* except occasionally in gross motor development such as walking He tends to be more backward in social behaviour and subsequently in speech than in other fields By social behaviour one means his interest in his surroundings the responsiveness to his mother including smiling his alertness and concentration (*e g* in the duration of time for which after the age of five or six months he will play with a given toy or try to get one out of reach) He tends to be relatively less retarded in motor development—the age of sitting and walking—and sometimes in sphincter control than in other fields The situation is rather different for the child who develops normally for a few

in an infant who is failing to thrive and perhaps suffering from vomiting polyuria and constipation It may well prove that other metabolic conditions associated with mental deficiency are amen able to treatment and every opportunity to study these cases increases the chance that something can eventually be done to help them

Apart from the above reasons for the importance of early diagnosis it is thoroughly desirable that parents should be told as soon as possible about a child s mental deficiency so that they may face the issues involved and plan the future Before they are told however the diagnosis must be certain No hint about the possibility of mental deficiency should be made while this is still in doubt

THE NORMAL DEVELOPMENT OF THE INFANT

The first step to the diagnosis of the abnormal is a thorough knowledge of the normal and of the variations from normal which occur in the absence of disease

In the section below it is assumed that the baby was born at term If he were born prematurely an appropriate addition must be made for his milestones of development For instance if a baby is born two months prematurely he will be expected to smile at the mother at the age of six weeks plus two months

The average six weeks old baby will smile at his mother when he is not crying and when she talks to him Prior to that he watches his mother intently as she speaks to him—provided he is not crying from hunger or otherwise uncomfortable When held face downwards with one s hand under his abdomen he is able to hold his head up at least momentarily in the same plane as the rest of the body

At three months of age he shows great interest in his surround ings He turns his head to follow a moving person with his eyes or to look for the source of a sound When placed on his abdomen he bears his weight on his forearms with the chest well off the couch He will hold and play with a rattle placed in his hand

questions in such a way that the mother understands exactly what they mean For instance the age of beginning to smile is an important milestone but it is necessary to be sure that the mother understands that one is referring to the age of smiling in response to her overtures Some mothers interpret any wince of pain from wind or facial movement in sleep as a smile The only information of value is the age at which the child first began to smile at the mother when she spoke to him when he was awake and not crying because of hunger or other reasons The age at which a baby first succeeded in going for an object *and getting it (average five months) is important the mother* must clearly distinguish this in her answer from the age at which he would hold a rattle when placed in the hand (average three months) When questioned about the age at which a child learnt to sit the mother must clearly understand that one is asking for the age at which the child first sat for a few seconds on the floor without falling over (average seven months) This is very different from the age at which the baby could sit up in the pram without a pillow supporting him but with support from the mattress around the buttocks (average five months) In asking about the age of walking the significant age is that of walking without any support (average thirteen months) and not the age of walking with hands held (nine months onwards) It is useless *to ask the mother when the child first talked She is likely to* interpret as speech the child s repetition of syllables (dadada bababa from seven months) whereas what one wants to know is the age at which he first spoke single words with meaning A child may repeat two or three words such as Oh dear in imitation of a parent but this is different from spontaneously joining words together (average twenty one to twenty four months) As for sphincter control the only information of value is the age at which the child first began to tell the mother when he wanted to use the pottie (average fifteen months) the age at which he subsequently became reliable by day (average two years) and later by night (average three years) The early conditioning to use the pottie when placed on it (from a week or two of age) is useless as a milestone The age at which the child began to feed himself is useful this can be defined as the age at which he could first be relied upon to pick up an average full

months and then as a result of encephalitis or some degenerative disease of the nervous system or other cause undergoes mental deterioration Such a child may seem at first to be relatively normal in the mechanical skills which he has learnt such as motor development but he is seriously defective in his social behaviour In particular he may show aimless overactivity with obviously defective concentration

It follows that the diagnosis of mental deficiency should never even be suspected if there is backwardness in one field of develop ment only such as motor development speech or sphincter control

The Value of the History

The family and perinatal history is of some value in diagnosis and a history of physical illnesses must be recorded because it may throw some light on the child s development but of much greater importance is the history of his actual development up to the present.

A history of mental deficiency in near relatives may be ob tained but little attention should be paid to it for some eighty per cent of children of a mentally defective parent are of normal intelligence It is obvious that the likelihood of a child of a mentally defective parent being himself defective is greater than it is in children of normal parents but the risk must not be exaggerated

A perinatal history of foetal distress anoxia at birth cyanotic attacks convulsions, undue drowsiness or even cerebral haemor rhage must be noted but once more their importance must not be exaggerated for the great majority of babies who have severe asphyxia at birth cyanotic attacks or convulsions grow up to be perfectly normal nevertheless the risk of an abnormality in such children is greater than it is in others

As for physical illness it is only serious chronic diseases which are likely to have any bearing on the child s development For instance if a baby were to be kept lying down in hospital from the age of four to six months one might reasonably expect delay in learning to sit up

The developmental history is of great importance in the assess ment of a child s intelligence It is essential however to ask one s

regurgitate and to fail to demand feeds like a normal baby It must be emphasized however that the great majority of babies who sleep excessively who regurgitate or who are drowsy in the newborn period and fail to demand feeds turn out to be mentally normal The slightly older baby—from one to four months or so—is often described by his mother as a good baby and not a bit of trouble because of his tendency to sleep more than other babies However not all defective babies sleep excessively

The main and all important sign of mental deficiency consists of backwardness in development On this and on this alone should mental deficiency be diagnosed The defective child is backward in all fields of development except occasionally in sitting and walking so that he is late in beginning to smile to vocalize as he smiles to follow with his eyes to turn his head to sound to use his hands and to chew Lateness in chewing often causes difficulties since his mother may attempt to give him solid foods because he is over six months of age and his inability to chew causes him to vomit He will usually be late in motor development, so that his head lags excessively when he is pulled to the sitting position in the first few weeks At six weeks instead of *holding his head in the same plane as the rest* of the body when held face downwards the head hangs down and the arms and legs hang down lifelessly whereas in a normal baby the hips are largely extended and the elbows flexed At three or four months of age he can only just if at all lift his chin off the couch in the prone position instead of holding his chest off the couch like a normal baby He is likely to be late in sitting creeping pulling himself up to the standing position and walking

More important than lateness in motor development is his lack of interest in his surroundings especially after the age of two months He lacks the alertness concentration and determination of the normal baby For instance when a normal baby of seven months drops a toy he will look to see where it has gone and he will try persistently to grasp a toy out of reach The mentally defective baby shows no interest in either

The normal baby can frequently be seen at the age of twelve to sixteen weeks lying on his back watching his own hand movements as he rotates his wrist and moves his fingers He should

cup drink from it and put it down again (average fifteen months) If the mother is not asked the question properly she will think it refers to the age at which the baby could take a biscuit to the mouth and eat it (average six months)

Properly taken the developmental history is of great importance Carelessly taken it means nothing and merely misleads

Teething is useless as a guide to development and has no relationship to the child s intelligence

Signs of Retardation

The signs of Mongolism and cretinism are well known and are adequately described in the textbooks of paediatrics It is worth noting however that the family doctor may first be led to suspect these conditions and in particular thyroid deficiency by detecting retardation in development

The so called stigmata of mental deficiency are quite unreliable and hardly help in diagnosis It is true that many mentally defective children and adults have anomalies of the ear and other congenital anomalies but these occur so commonly in the absence of mental deficiency that they are of no value in diagnosis

It is a great mistake to diagnose mental deficiency because the child has a peculiar face or head He may take after his otherwise normal father or mother in that respect Mental deficiency should not be diagnosed because the head is large or even if definite hydrocephalus is found a child may be born with a non progressive hydrocephalus and retain normal intelligence Neither should it be diagnosed because the head is unusually small Several famous people such as Lord Byron had unusually small heads It is true that very many children who are mentally defective from birth have microcephaly This refers more to the shape of the head than to its size and even microcephaly can occasionally be associated with a normal level of intelligence The microcephalic head tapers off unduly towards the vertex and the forehead slopes backward excessively

In the newborn period there are no definite signs of mental deficiency unless the child is a cretin or mongol It is commonly found however that a mentally defective baby sleeps excessively in the first few weeks and is more liable to feeding difficulties in the first week or two having in particular a tendency to

ment reached by the time he is seen and then he develops signs of a degenerative disease The commonest of these is the appear ance of the so called akinetic seizures or salaam spasms in which the baby has repeated sudden momentary forward jerks They have often wrongly been termed *petit mal* They are associated with severe mental deficiency The child very soon stops taking an interest in his surroundings stops smiling and becomes severely defective and yet owing to previous normal development he has learnt to sit or walk There are many familial and other degenerative diseases of the nervous system which are associated with progressive mental deterioration

DIFFERENTIAL DIAGNOSIS

Retardation in one field of development such as motor development speech or sphincter control should never suggest mental deficiency for the mentally defective child is never back ward in just one field he is backward in all except occasionally motor development and sphincter control It should be particu larly noted that backwardness in individual fields can occur in normal children without any disease being present For instance I have seen an otherwise normal child who was unable to sit without help until twenty one months or to walk without help until thirty months His I Q at five years was 102 and there was no physical defect Lateness in speech is common without any physical defect such as deafness even in children of average or superior intelligence Lateness in the acquisition of sphincter control is also common without any apparent physical cause In some of these varied forms of backwardness there is a family history of the same complaint

Backwardness in motor development may also be due to a disturbance of muscle tone such as hypotonia of which amyo tonia congenita is one of the well known examples or hypertonia as in cerebral palsy In the former case the very poor muscle tone is readily detected and the child shows a grave backwardness in gross motor and manipulative development yet the striking feature is that his social behaviour—the age of smiling and vocalizing the interest concentration and alertness which he shows—is normal immediately excluding mental deficiency

stop doing this after about sixteen weeks but a mentally defective child can often be seen doing it at seven or eight months or even later

Normal babies usually stop taking objects to the mouth after twelve months of age and largely stop slobbering except when they are particularly interested in a new toy or some other exciting object The mentally defective child continues both these habits long after this often till three or four years of age or more Babies of eleven or twelve months characteristically throw objects on to the floor they stop doing this in three or four months but mentally defective children may continue to do it for some years

The defective child is especially likely to be late in learning to speak so that no words are spoken with meaning until well after the first birthday and words are not put together until after the second birthday He makes no attempt to hold his arm out for clothes at the usual time (twelve months) Sphincter control is usually but not always delayed in these children They are all late in learning to feed and dress themselves though one must remember that both these skills can be greatly delayed by a mother s failure to give them a chance to learn

When a normal baby of six to nine months or so lies on the back the reciprocal kick can be seen—the kicking of first one leg and then the other This disappears (except in marked pleasure or displeasure) when he learns to walk it continues for an undue period of time in the defective child

After the defective child has begun to walk he may show aimless overactivity and destructiveness He rushes round a room picking up first one object and then another with no sustained interest in anything

DEGENERATIVE DISEASES OF THE NERVOUS SYSTEM

The various degenerative diseases of the nervous system occupy a special place in a discussion of early diagnosis because their features differ from those of simple mental deficiency In these the child develops normally for several weeks or months as shown by the developmental history and the stage of develop

spasm of the thigh muscles is tested for by flexing the hip to a right angle and then rapidly abducting the hip Once more the interpretation of the test is a matter of experience except in severely spastic babies

The persistence of primitive reflexes is of some value in the diagnosis of cerebral palsy The grasp reflex and tonic neck reflex may persist long after three months when they are normally lost The grasp reflex is tested by slipping the finger into the palm of the baby s hand when he closes on the finger and one can lift him off the couch by it The tonic neck reflex is seen when the baby is lying on his back he may extend one arm to the side to which his neck is rotated the contralateral knee being flexed

The *athetoid form* of cerebral palsy cannot be diagnosed until the characteristic movements are detected and this may not be until the age of three years or sometimes much later It should be suspected in any retarded baby with a history of severe asphyxia at birth inadequately treated haemolytic disease of the newborn or a history of improperly treated severe jaundice in the newborn period associated with prematurity These latter two conditions are apt to lead to kernicterus of which athetosis is a major sign

In an older child (*e g* after the first birthday) the speech development is of great importance in assessing the intelligence in a case of cerebral palsy and so in eliminating severe mental deficiency *If for instance a child aged eighteen months is saying* several words with meaning and yet is very far from walking and perhaps cannot even sit he cannot be severely mentally defective for if he were his speech would not be nearly so advanced In *mental deficiency speech is relatively more retarded* than motor development.

Sensory defects involving hearing and sight may cause the unwary to diagnose mental deficiency because of the effect on speech in the case of the former and on social responsiveness and manipulative behaviour in the case of the latter In other fields of development however the child will be normal

Considerable general retardation may be caused by severe chronic illness though the history of this will be readily obtained It may also be caused by severe chronic emotional deprivation

The diagnosis of **cerebral palsy** is more difficult because it is so frequently associated with other defects and in particular with mental deficiency It has been shown that more than half of all cases of cerebral palsy have an intelligence quotient of less than seventy and this in itself has a retarding influence on general development Cerebral palsy is common—affecting about 1 5 per 1 000 school children—and it follows that whenever motor or general retardation is found cerebral palsy must be suspected and looked for The first sign of cerebral palsy is delayed motor development In the case of a six weeks old child for instance the first pointer to the presence of cerebral palsy is likely to be excessive head lag when the child is held face downward When therefore backwardness is found the first step is to try to decide whether this backwardness is confined to one field or applies to all if it is a general backwardness the commonest cause is mental deficiency The second commonest cause is mental deficiency plus cerebral palsy If the child has cerebral palsy alone with a normal level of intelligence he may be very backward in motor development but he will be interested and alert and will follow well with his eyes and concentrate well (*e g* on trying to reach toys after the age of six months) If the level of intelligence is low as it usually is the child will be backward also in this social field

The *spastic form* of cerebral palsy constituting seventy per cent of all cases is diagnosed by the detection of exaggeration of the knee jerks and possibly by the finding of ankle clonus and of adductor spasm in the thigh muscles In addition after the age of three months one hand may be found to be persistently closed in the case of a hemiplegia whereas it should be open at that time or both hands may be closed in the case of quadriplegia The knee jerks may be tested by tapping over the front of the ankle for when disease of the pyramidal tract is present the area over which the knee jerk is obtained is greatly increased If therefore a brisk response is obtained when the front of the ankle is tapped it is likely that disease of the pyramidal tracts is present It is obvious that interpretation of the response requires experience

Ankle clonus is tested for by gentle but rapid dorsiflexion of the ankle by one finger under the side of the foot Adductor

investigations are only of value to exclude a few rare types of mental deficiency

SUMMARY

The early diagnosis of mental deficiency depends on a thorough knowledge of the normal and thus on the recognition of retardation in development Except in the case of cretinism and mongolism it does not depend on the facial appearance

The mentally defective child is backward in all fields of development except occasionally in gross motor development and in sphincter control he is particularly backward in social behaviour and in his social responsiveness his interest in his surroundings his alertness and his power of concentration Speech is relatively more retarded in mentally defective children than other fields of development It follows that mental defectiveness should not even be suspected if retardation is found in a single field such as motor development speech or sphincter control

When severe general retardation is found in an infant the commonest cause is mental deficiency but the second commonest cause is a combination of cerebral palsy and mental deficiency Thus in all babies in whom general retardation is found cerebral palsy must be looked for

A careful history is the basis of diagnosis which depends also on an informed developmental examination and the interpretation of the combined findings Special investigations are of limited value

as occurs when a child is brought up in an institution from birth Such children are retarded in motor and social development in speech and often in sphincter control

Very occasionally a child is seen who is severely retarded in motor development in the early weeks or still more rarely in the whole of development and who subsequently over the course of the first year catches up to the average and becomes a normal child without any physical or mental handicap These cases are rare but they must be recognized Some may result from birth injury others are unexplained It follows that care must be taken not to give a confident opinion about a child in the first few weeks though these special cases are so rare that on finding severe uniform retardation in a young baby one will only very occasionally be wrong in suspecting mental retardation with or without cerebral palsy

Not infrequently it is found that development in one particular field especially speech seems to be at a standstill for several weeks without any apparent reason This is a normal pheno menon and must not lead one to suspect mental deterioration Such children are clearly developing normally in other fields

THE USE OF SPECIAL INVESTIGATIONS

Special investigations are of little use in the early diagnosis of mental deficiency except in the confirmation of a diagnosis of thyroid deficiency In a cretin the total serum lipoids are raised and X rays show retardation of bone age These are invaluable investigations in a doubtful case No special investigation is of value in the diagnosis of mongolism which is based chiefly on the facial appearance An electroencephalogram is of value in inter preting the type of epilepsy from which a child is suffering and in the diagnosis of certain forms of encephalitis and degenerative diseases of the nervous system Examination of the cerebrospinal fluid and more specialized neurological investigations are of value in certain rare types A Wassermann reaction is useful in elimin ating a syphilitic infection in certain cases If ophthalmoscopic examination reveals a choroidoretinitis an X ray of the skull should be taken and skin and blood tests should be done for the diagnosis of toxoplasmosis In general however laboratory

The latter if he takes these half intended hints and acts firmly may undoubtedly prevent the onset of addiction He may on the other hand lose his patient He will in any case get little thanks even if he is successful for the patient helped in time is not likely to admit to himself that he might have gone on to alcoholism None the less the practitioner who has known the patient over the years as well as his family and social background and who has seen the change in him is in a unique position to help It is essential therefore that he should know how to identify the point at which drinking becomes something more When the diagnosis is made too it must be sufficiently confident to survive the patient s protests and explanations and to carry conviction when it is put to him The treatment of addiction in general is perhaps exceptional in that it is better for the patient that his physician should feel as well as convey an unwavering certainty as to diagnosis

For the purpose of this article alcoholism is a disorder of multiple aetiology characterized by drinking which has gone beyond the dietary use of alcohol and the extremes of compliance with the drinking customs of the community and is associated with impairment of personal efficiency and relations with others Alcoholism is present when the drinker has reached a point at which his own resources are insufficient to prevent a progressive addiction leading to serious changes in personality and irreversible damage to the nervous system and other organs

THE SPECIAL DIFFICULTIES OF DIAGNOSIS

The wide range of vulnerability to alcohol in different individuals is well known whether the causes of this be in inheritance early influences physical disability or personal misfortune In some cases it can be seen retrospectively that the point of no return was virtually at the first drink taken whereas in others addiction is established only after years of heavy drinking in an occupational or social milieu in which this is part of the accepted mode of life There is also wide variation in the physical *or psychological function which is the first to show signs* of failure In some it is the liver in others the peripheral nerves while in yet others it may be memory or the sense of responsi

CHAPTER XIX

ALCOHOLISM

BY ALEXANDER KENNEDY

THAT one can lead an alcoholic to the doctor but cannot stop him drinking is too often the bitter experience of those who have tried to help one or have become involved in his affairs In most cases by the time he is referred for specialist opinion the diagnosis is plain for all to see By then the main problem lies in installing in the patient a will to recover that will last more than a few brief hours

Nevertheless between the taking of alcohol within the conventions of our society and serious addiction with breakdown there is often a time when the excessive drinker is approaching the point of no return and it is then that an early diagnosis is of the greatest value At this time the shallow self deceptions and the extraordinarily specific lack of foresight in matters alcoholic of the confirmed addict are not yet installed The patient may have been horrified suddenly to discover that he cannot give up alcohol at will It is at this time that he may send out half hearted and ambiguous distress signals to his non drinking friends and to any doctor whom he meets socially He will consult his general practitioner about gastric symptoms loss of sleep or the effects of overwork and will describe symptoms suggestive of anxiety or depression in the ambivalent hope that he will be asked questions about his drinking habits and have his resolution fortified by the recommendation of a period of abstinence on medical grounds A history of several months abstinence imposed on account of symptoms resembling those of peptic ulcer is very common in the early stages and the fully addicted drinker may later point to this piece of ancient history with pride as evidence of his ability to stop if he really has a good reason It is at this point that the potential alcoholic is apt to make joking references to his dilemma or to put it to his doctor as a hypothetical case

the house At any point the alcoholic may terminate the proceed ings by declaring that the questions are going beyond legitimate medical enquiry and by taking offence on the ground that he is being suspected of lying That this does not in fact happen more often is due to the presence of an unconscious desire to confess and the doctor must keep this to the fore by mentioning the advantages of having the aid of someone else in dealing with the problem

In most patients the fear of humiliation looms especially large because the alcoholic has usually covered his inward fears by outward talk of his capacity to hold his liquor his ability to afford to drink as much as he likes and his generally manly attitude to his personal problems For this reason it is essential that no element of judgement or moral censure should enter into the approach to these patients When statistics which are obviously false are offered as to the amount taken or as to the effect of drinking on his work or mental happiness it is better to confine oneself to mental reservations until a number of statements have been made which can be challenged by facts that cannot be explained away There are often moments when for a short period of morbid drunkenness or post alcoholic remorse a train of confession can be started and it is then that the facts come out Such periods are often brief however and it is by no means uncommon for an alcoholic to summon his doctor as a matter of urgency with the intention of telling everything only to recover his self confidence and revile him jovially when he arrives

In order to establish a diagnosis it is necessary therefore to steer a way through these difficulties and to tolerate the evasive ness the idealistic pretensions the unscrupulousness and egocentri city the self justifications and the cliches which have gradually replaced normal responses in a man to whom drinking has been at first an abiding interest and later an inescapable burden The physician must listen to the defamation of those who have tried to help the patient before the solemn but empty promises the wife who is taken for granted (grand girl) and the use of phrases which *he knows will invariably precede a major pre* varication such as to be quite frank with you honestly or as man to man

bility which proves to be the weakest point. The clinical development of alcoholism is thus seen to be a very individual matter both in point of symptoms and the order of their appearance. In spite of this and of the variety of patterns of dangerous drinking there are features common to almost all alcoholics even in the earliest stages and as the disorder progresses the patient tends to lose his independence as an individual personality and to conform in most respects to a predictable pattern It is these general features which as they make their appearance one by one render diagnosis progressively more certain

When taking a history with the suspicion of alcoholism in mind the physician is in the position of one seeking to bring to light information which if he is successful will lead to the embarrassment and discomfiture of the patient who well knows that he will be called upon to give up alcohol He will if the diagnosis is brought home to him have to admit defeat by an enemy that he has scorned for long enough It is characteristic of the addict that this immediate fear of humiliation is more prominent in his mind than consideration of the benefit he will get by seeking medical aid He has averted his attention so long from the facts of the situation that anyone who attempts to make him face them will encounter a variety of mental mechanisms of defence. Of these the main ones are the *suppression of essential details* the *minimization of important facts* such as the amount of drink taken the *facile explanation* of behaviour with which he is taxed and a *superficial and light hearted attitude* to the subject with *appeal to conventional ideas* about alcoholic excess and *comparison of his case with that of others* who are said to be drinking at least as much with impunity Medical inquiry has the effect of accentuating the alcoholic s inward conflict between admitting defeat and seeking help on the one hand and attempting to continue his efforts to deal with the problem alone on the other When in these circumstances he tells a direct lie he is also deceiving himself for his conflict has brought about a neurotic reaction in which his memory of unpleasant events is momentarily suppressed In the case of some female secret drinkers the physician will have to contend with a facility in falsehood which can stand up to any medical interrogation and even to the production of circumstantial evidence in the form of bottles concealed all over

organizes his life mainly round the drinking situation to the neglect of other interests Indeed it is estimated that only one twentieth of regular heavy drinkers eventually become addicted If abstinence is suggested on medical grounds the heavy drinker can stop without difficulty and resume when the prescribed period is over He drinks because he likes it and sets an accurate limit to his capacity to which he adheres on all but special occasions He drinks at an even pace rarely drinks alone eats adequate meals and is not distressed by a few hours abstinence from alcohol He has chosen this way of life and within its limitations is well adjusted to it

At the same bar is the early alcoholic and his actual intake may be less It has however to be considered in relation to the vulnerability of his personality After a varying time he separates himself off from the rest by the development one by one of certain characteristic features

When with friends and acquaintances he draws them to the bar as if it were the only possible place to go He gulps down the first two drinks of the evening and rather forces the pace even having drinks between rounds He regards a period of drinking before meals as a ritual never to be omitted and embarrasses others by putting off going in to dinner until he has had enough He holds his liquor well at this stage and regards all events as calling for a drink He spends an increasing amount of time away from home but refers to his family in a rather sentimental way

When travelling he avoids hotels trains and planes without bars and is happiest at an international airport with low priced drinks and no closing hours He often miscalculates the amount of currency he needs to carry and may have to borrow When settling down for a journey he looks for travelling companions who will join him in a drink He carries a large flask by way of reserve and on return to this country regularly brings a couple of opened bottles of spirits through the customs

When going to a party he often takes a drink before starting out and on arrival makes a quick start so that he can feel sociable He is without shame in scorning the cup and going for the whisky which the hostess holds in reserve and the length of his stay depends on the availability of drink He may at this

As the indirect evidence provided by these evasions gathers strength it is wise to obtain some direct observations from other people especially before direct and difficult questions are put to the patient The alcoholic is often very loth to allow his wife to see the doctor and if afterwards he feels she has let him down he may behave very badly when he gets home It may be difficult to get the facts on account also of a misplaced loyalty for it seems that nothing brings out the qualities of female generosity and forgiveness more than an alcoholic husband None the less the wife should be seen if possible and it may be necessary to wait for the alcoholic to protest the truth of his statements by saying You can ask my wife you can ask them at work if you like and then to obtain his permission to do this Little can be learned from those with whom the patient drinks as one then often comes up against the group defensive reaction instead of the individual one It is noteworthy that by the time a female alcoholic has reached the confirmed stage the husband unless he is an alcoholic himself has either left her or is proclaiming that he will do so if nothing can be done When it is the husband or wife of the patient who comes first there is usually abundant evidence and the difficulty is to confirm it by reference to the patient without increasing the antagonism between the two When both partners are alcoholic the defensive teamwork in the consulting room is often in striking contrast with the altercations and disharmony of the home

It will be seen that it may be difficult to help an alcoholic who is afraid to be helped If his co operation is finally obtained the doctor then acquires a difficult and exacting patient from whom no sort of gratitude is to be expected The temptation to allow the patient to lie his way out and to regard the matter as coming within the moral rather than the medical sphere is often strong It is however a test of the physician s sense of responsibility to prevent disease at the stage when it can be prevented

PREMONITORY SIGNS OF ADDICTION
(Stage I)

The regular taking of large quantities of alcohol does not necessarily imply a diagnosis of alcoholism even if the drinker

occasional drinkers after a heavy blind but their repeated occurrence after a relatively small intake is a danger signal Like the boxer who experiences similar blackouts and who must give up the game or become punch drunk the alcoholic must stop at this crucial point before his capacity to resist addiction unaided slips away

PREVIOUS PERSONALITY—THE SYMPTOMATIC DRINKER

Although at the stage of final breakdown alcoholics tend to conform to a common clinical pattern there is no such thing as a characteristic pre alcoholic personality The reasons why each starts to drink immoderately differ greatly and the combination of constitutional vulnerability and the stresses and circumstances of life will determine whether he will go on to addiction Indeed in retrospect it is sometimes evident that the patient could hardly have done otherwise than take to or be driven to drink In some families where alcoholism is almost the rule and in which psycho pathic tendencies are common the constitutional factor is almost overwhelming while in the case of those of high intelligence who have been deprived of outlet or opportunity or in the husbands of psychotic wives who devote their lives to persecution it has been the obvious escape

There are however many instances where resort to alcohol occurred on account of circumstances and states of mind which had they been recognized in time could have been alleviated before the onset of addiction These symptomatic drinkers are the patients in whom early diagnosis may really lead to a return to normal life with relatively small prospect of relapse

Amongst these the patient who can most readily be helped is the recurrent depressive who has used alcohol as a symptomatic treatment for his endogenous unhappiness In the early stages the periodicity of the drinking the frequent family history of manic depressive disorder and the history of swings of mood before the onset of drinking will often make the diagnosis plain Treatment of the depression or even explanation of the nature of the illness and the fact that recovery is bound to occur will usually enable the patient to stop As in any other drinker who

stage joke about alcoholism and refer to himself as an alcoholic' or a fellow sufferer

At work he believes that a drink is essential to the transaction of business goes out to lunch early expects to be covered by his colleagues and talks about the strain of modern business life He tends to be late and uncommunicative in the mornings He does not entrust his expense account to his secretary but makes it up personally

At home he takes the support and help of his wife for granted and if she has some money of her own he will invest it for her Unless he is very well off he is a strong supporter of state education and does not pay private school fees or send his children to the University If asked about this an early start in business' and the University of Hard Knocks is his preference for them He conceals his exact income from his wife and a call for housekeeping money may lead to a scene which ends in his going out for a drink At times however he is affectionate with bursts of generosity towards the children presents and promises to be at home more in future

When alone he may drink to relieve fatigue or tension He may later drink a hair of the dog that bit him in the mornings He becomes aware of the amount he is drinking only when he finds his wallet empty At intervals he worries about his drinking and sets himself target amounts makes promises or resolves to drink only certain drinks at certain times

In the consulting room he appears with symptoms of a general kind attributing them to overwork—or to anything but alcohol There may also be gastric symptoms When asked about alcohol he will often admit that he had taken too much at times although his statistics are unreliable He will be confident of his capacity to regulate his drinking and may give effusive thanks to his physician for having made him realize the situation If these opportunities are not followed up he will usually relapse He may soon tire of his doctor's efforts and think of him as nagging as having a bee in his bonnet about alcohol or as exceeding his duty of relieving the original symptom

There is one symptom of special importance which often marks the transition to confirmed alcoholism and that is the occurrence of blackouts or periods of amnesia These may occur in

watched when they make their painful readjustment to life lest they side step the effort involved and retreat into alcoholic solace A specially important instance is the elderly widow who is suddenly alone and who begins to drink secretly almost from the day of the funeral In two of the writer s patients the drink taken on that occasion was virtually the first experience of the comforts of alcohol Another isolated type is the lonely male homosexual of good behaviour whose recourse to alcohol may later lead to release of the underlying trend

Amongst the inhibited who cannot meet other people without a drink there are many who can be helped Of these a special group are the sexually inhibited and there are instances of married couples who have from the first only been able to enjoy intercourse with the aid of alcohol and who have gradually evolved a drunken partnership The prostitute who can only face her occupation with the aid of alcohol must be reclaimed early if there is to be any chance of permanent rehabilitation

Opportunity of obtaining alcohol at minimal or no cost with the leisure to drink it leads sometimes to addiction amongst workers in the drink trade barmen and barmaids as well as ship s medical officers while the rich and idle may similarly take too much at a time of relatively minor stress A group which has recently become important is the domestic makers of rustic wines who may accumulate large stocks of elderberry or dandelion wine and devote themselves to solitary sampling A final symptomatic drinker is the diabetic of slow onset who drinks to relieve his thirst A pathetic example of this was the son of an eastern ruler who excused himself when the diagnosis had finally been brought home to him by saying No one would think of bringing me a Prince a mere glass of lemonade

PATTERNS OF DRINKING

Alcoholic drinks are taken in a variety of settings which vary with local and national social or business usage The attitude to drinking varies from the family who without being nominal abstainers think they are going far enough with cider or ginger wine on Christmas day to the alcoholic who goes on benders for a fortnight at a time with little interval between Some

has been in danger of addiction *lifelong abstinence thereafter* has been shown to be essential Another group who use alcohol as a drug for treating their symptoms are patients with chronic anxiety states Among these have been some remarkable instances of the constitutionally anxious type with a determination to conquer fear Some of these after outstanding war service have developed alcoholism in time of peace Where the anxiety is due to psychological conflict an appraisal and readjustment may relieve the anxiety while in others more conventional symptomatic treatment with barbiturates or ataractics may relieve the tension in time

A special group of alcoholics characterized usually by periodic drinking are the dysrhythmics whose cerebral activity is the subject of storms which are the equivalents of epilepsy Here the patient subject to unpredictable tension and irritability has found his own anticonvulsant Diagnosis can be made with the aid of the electroencephalograph and less dangerous anticonvulsants substituted The diagnosis is not uncommonly made plain by the appearance of a fit at the end of a bout of drinking or by an amnesia or outburst of violence

In the case of the inadequate psychopathic personality without *drive or capacity to find a stable place in the community* and born several drinks under par the material is poorer and even if suitable work and alternative interests can be found the danger of relapse is much greater When however the inadequacy is relative as in the not uncommon instance of the man who has inherited the direction of a family business which is beyond his intelligence or remote from his interests an adjustment of the underlying situation will make the task of giving up alcohol much easier Rather more difficult is the man of high intelligence who must work and live with people whose interests are superficial and minds limited He is found to be literally trying to drink his I Q down and a major reorganization of his life *may be necessary if he is to recover*

In many instances especially in those who take to drink relatively late in life alcohol serves as a form of neurotic escape The newly blinded or seriously disfigured must be carefully

watched when they make their painful readjustment to life lest they side step the effort involved and retreat into alcoholic solace A specially important instance is the elderly widow who is suddenly alone and who begins to drink secretly almost from the day of the funeral In two of the writer s patients the drink taken on that occasion was virtually the first experience of the comforts of alcohol Another isolated type is the lonely male homosexual of good behaviour whose recourse to alcohol may later lead to release of the underlying trend

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patterns are almost entirely benign so far as alcoholism is concerned and include the drinking of wine with meals the occasional cocktail party the regular drink before dinner or the discussion of port after it. The great majority of social drinkers or those who confine their drinking to the business lunch or club dinner are in no danger On the borderline is the public relations man or salesman whose thirst is not quite satisfied within his expense account and the public house regular who confines himself to beer The man who has a drink on most days and overdoes it on an occasional spree is rather less under control and contrary to general opinion the amateur of wines is not always able to restrict his subjective investigations to an appraisal of colour bouquet taste and vinosity The heavy beer drinker often one whose work is hot dry damp or just thirsty can usually gauge his capacity as can the gregarious habitual drinker of spirits to whom the permitted drinking hours are the best part of life From this point however the patterns of drinking are associated with addiction more often than not *The periodic* drinker may be of cyclothymic personality but he may come to drink not only in his depressive phases but when he is under any kind of stress Some of this group especially those with an epileptic background merit the term dipsomaniac the compulsive drinker who once started cannot stop and who drinks himself to extinction with decreasing intervals between each bout Virtually all secret drinkers are addicts Secret drinking is commoner in women and if one stands at an off licence counter for a while the procession of poorly dressed women with shopping bags who come in and place their money on the counter without comment receive a bottle of port type and rather furtively depart indicate that secret drinking is not confined to those who can well afford it Those who can usually place a regular order with two or more suppliers often grocers who know that to deliver without fuss is to retain a valuable customer The bottles are taken in without the case in which they were brought and distributed about the household so that no single cache contains more than a reasonable amount. The account is incorporated in the monthly groceries bill and the husband is apt to feel strongly about inflation and the rising cost of living A search conducted after a patient of the writer had been admitted to hospital for investiga

tion led to the discovery of over three hundred gin bottles in differing degrees of emptiness It transpired that the lady of the house took a little here and there as she teetered round her domain Amongst such secret drinkers are found the amoral psychopathic women with a lurid history of love affairs who take to alcohol to assuage an increasing sexual interest with the onset of middle age In these the prognosis is usually poor There is one pattern which may be considered more as a classical sign of alcoholism than a pattern of drinking and that is the occurrence of benders These are bouts of drinking in which work and all other obligations are forgotten for days at a time and which end in complete exhaustion often with severe remorse At first the bender drinker is able by a remarkable and characteristic effort to get himself back to work when it is over but later the periods of drinking fuse as withdrawal symptoms drive the patient to seek relief in more alcohol

The majority of regular users of alcohol although disinhibited and able to repress current worries are able to keep within an accepted standard of behaviour In others smaller amounts of alcohol produce changes in behaviour which make them conspicuous even in drinking company The subjects of this pathological intoxication include the amorous the aggressive the sexually inverted and the passer out In spite of regretting what has happened these tend to become addicted very easily and if they can be persuaded after getting into trouble that they should leave alcohol alone altogether alcoholism may thereby be prevented A curious form of pathological drinker is the individual who when intoxicated insists on recounting the less favourable aspects of his past in a sort of cathartic session addressed to anyone who will act as analyst Related to this type is the alcoholic who drinks heavily at intervals and then makes a *serious attempt at suicide Such drinkers can be regarded* as chronic suicides and their condition is one of the few indications for advising the police to charge anyone with attempted suicide Only in this way may the inevitable outcome be prevented by securing a short sentence in the Magistrates Court and treatment in the prison hospital

THE ALCOHOLIC'S PROGRESS

The earliest stages of addiction have already been described Once this is established there is a fairly uniform progress differing from one patient to another more in its time relations than its clinical features Three further arbitrary stages will now be described

The Established Alcoholic (Stage II)

By the time this stage has been reached a day is rarely spent without some drinking although it is still possible to stop drinking for a short period when necessary Unlike the regular and heavy but unaddicted drinker who can abstain if so advised for medical reasons for months at a time the alcoholic starts once more against his own inclination usually after only a few days By this time he only feels at ease if he has a glass in front of him He still has a good head and can often pull himself up into a state of apparent sobriety if for instance his car is stopped by the police or if he meets someone with whom he must be wary if he is to keep his job He is beginning now however to lose the ability to regulate the quantity he takes and his first drink creates an immediate desire for more until a comfortable concentration is reached He is therefore apt early in the evening to take drinks between rounds and even to excuse himself and go out and have a nip from the bottle in his car Short periods of remorse now make their appearance and in these he will make solemn promises to his wife or friends It is at this stage that the financial effects of drinking usually begin to be felt and betting is common as well as uncritical entry upon schemes for getting rich quickly If in a position of trust he may now accept presents in exchange for favours or for not noticing irregularities His worries about money can be set aside while he is drinking but are apt to come out in the form of boasting about his ability to look after himself in this respect and carrying five pound notes for display when he opens his wallet to pay for the round of drinks He has difficulty in remembering the details of his expenditure and his attempts at accounting leave him puzzled and uneasy There are gaps in his memory for some of his evenings although he would rather improvise than admit

this His attitude to his domestic responsibilities is by now entirely unsatisfactory and for this shallow excuses are made When in a public house he will subscribe to collections for the blind spastics dumb friends and similar charities with a momentary tear of sentiment but is becoming very cautious about buying drinks for other people or parting with money to his wife His attitude to the unfortunate even the other alcoholic is of the pull up the ladder Jack I m all right kind His family are referred to in cliches wonderful mother best set of kids in the world but he spends less and less time with them His sexual drive usually expires completely By this time he has lost all other interests but he is beginning to dislike talk about alcohol He is missing quite a serious amount of time from his work and his troubles may be aggravated either by a fall in his earnings on commission or by dismissal

The Advanced Chronic Alcoholic (Stage III)

By the time this stage has been reached severe spells of maudlin remorse are occurring frequently and the mention of drinking if he is not sufficiently drunk at the time reminds the alcoholic of his predicament He is constantly preoccupied with the fear of being without alcohol and lays in hidden supplies He is neglecting his food and stays away from home as his drinking acquires a compulsive quality characterized by bouts of several days duration He will by now have to drink to obtain sleep and to get himself going in the mornings and also to overcome the gross tremor which may necessitate his tying a towel to his arm and round his neck to steady him when he has to sign his name He has drifted away from his former drinking friends who although he excites pity in them find that his presence makes them uneasy and that it interrupts their routine He drifts from one public house or hotel to another muddles his way into parties where he is a complete stranger and causes embarrassment with his bouts of self pity and shallow excuses made in a slurred and *mournful voice He is still only willing to admit* defeat even to himself for short periods at a time By now his breath and body have acquired the peculiar odour of one whose metabolism has come to depend very largely on dilute ethyl alcohol

The Stage of Alcoholic Breakdown (Stage IV)

One of the first signs of the end is the fact that it takes less to make the alcoholic intoxicated than formerly He now is and looks drunk most of the day If he is not to find himself alone he may have to accept the company of vagrants and *drifting misfits and he may have to sleep rough or go to* the lowest type of lodging house On prolonged benders he may find it necessary to drink red biddy' or other forms of cheap liquor or even methylated spirit or the supernatant liquor of brass polish His morning nausea is now accompanied by vomiting so that he cannot always hold down his first morning drink Without alcohol his sense of weakness is such that he is unable almost to move and he is completely without initiative except to get more His ethical deterioration is almost complete and if he finds himself without drink he will steal often from the houses of friends who have tried to help him He may pay for drink with stolen cheques or cheques upon his own large overdraft His mental horizon is for the immediate future only and his memory of the recent past uncertain His isolation worries him and he will fear to walk in the dark alone At intervals he will admit defeat and it is characteristic of practically all alcoholics who reach this stage that they will make an attempt at prayer either to the Deity of their schooldays or in a more general To whom it may concern manner Fear intense nausea and exhaustion finally combine to produce for the first time an aversion to alcohol From this point it is a matter of no more than two or three days to the onset of delirium tremens

A SYSTEM OF EARLY DIAGNOSIS

It is not at all uncommon if the patient happens to be in a remorseful mood or if he is willing to admit his condition when persuaded to seek medical advice by friends for the diagnosis to be made from a history given by the patient Often however although it is strongly suspected the diagnosis cannot be made with sufficient certainty to advise the wholesale disruption of life which treatment involves Diagnosis can however be arrived at by a system of increasing probability if *the following factors* are considered —

Aetiological Probability

If there is a strong family history of alcoholism or if the patient s occupation is one in which alcoholism as opposed to heavy drinking is common Regular army officers whose rank is junior for their age senior N C O s repatriated tea planters public relations men motor salesmen the second and third generation of families in the drink trade and dentists who hate their occupation are the kind of people in whom there is a greatly increased probability

Personality Change

If some idea is obtained of the kind of man the patient was in his twenties and of the general trend of his career and ambitions comparison with his present position may help It must be remembered however that other types of organic deterioration of the nervous system may produce this effect though much more quickly as a rule

From the Testimony of Wife or Friends

If she is persuaded of the need for some kind of action the wife will tell about her husband s absences his drunken condition and his statements made in periods of remorse She will also be able to give an account of the effects on the family budget Alcoholism not infrequently comes to light through the discovery of financial irregularities and the patient may then if his firm are willing to close the matter accept treatment as a medical excuse

Circumstantial Evidence

The purchase of large quantities and the concealment of bottles may tell its own story as may the discovery of letters from the Bank or from the employer indicating serious neglect of normal responsibilities

From the Patient s Method of Evasion

The way in which he avoids the issue makes light of it understates his consumption of alcohol and glosses over his failures is quite characteristic and since it differs entirely from the way

a heavy non addicted drinker gives an account of himself is very strong diagnostic evidence

By the Presence of Physical Disease

The presence of hepatic cirrhosis chronic pharyngitis and conjunctivitis or achlorhydric gastritis will at least indicate a heavy alcoholic intake while dilated peripheral vessels the characteristic grog blossom' on the face and the sickly sweet smell of the heavy drinker may be inconsistent with the patient s account Polyneuritis may be present quite unknown to the patient

From the Patient's Mental Status

No profound knowledge of psychiatric techniques is necessary to detect the loss of recent memory the coarse tremor of the hands the superficial approach to all subjects the emotional facility the replacement of all original thought by set phrases and cliches and the tendency to cover the same autobiographical ground several times By the time a Korsakow s syndrome is present or the confusion of an alcoholic delirium the diagnosis will be obvious on other grounds

Confirmation of Diagnosis—the Test of Abstinence

Once the diagnosis is fairly certain the best way of confirming it is to impose a short period of abstinence by admitting the patient to a hospital or nursing home Precautions must be taken to relieve him of the supplies he will bring with him It is almost impossible to deal with him at home for this reason Abstinence symptoms will make their appearance within forty eight hours in the established alcoholic and may be used as a means of convincing the patient that he is in need of help Since the milder abstinence syndrome responds rapidly to treatment this must be withheld until the patient has seen how dependent he is on alcohol The withdrawal syndrome as seen in the state of alcoholic breakdown is in fact the prodrome of delirium tremens and has no relevance for early diagnosis The milder effects seen in the earlier alcoholic are however of great diagnostic value

The Withdrawal Syndrome

This begins with a feeling of restless tension and an over whelming desire to find a drink There is great irritability and sleep is interrupted with nightmares The patient worries about his physical health mentioning symptoms unrelated to his real disability He may become unexpectedly tearful and a childish dependence on the physician may alternate with a sudden deter mination to leave Tremor of the hands and arms is obvious and there is a sense of weakness with inertia which is the reason the patient usually does not go home Appetite is almost absent and severe nausea is present in the mornings There are attacks of vertigo and profuse cold sweats These are often accompanied by diarrhoea Rapid relief is obtained by giving alcohol chloral hydrate or paraldehyde The duration of this early withdrawal syndrome is from twenty four to seventy two hours and it can be terminated by giving thiamin by injection (100 mg) with 200 mg of nicotinic acid by mouth in four separate doses with a good deal of water Symptomatic relief is best secured by the use of chlorpromazine in 50 mg doses four hourly at first

The final confirmation must come from the patient himself He must realize that there is no doubt whatever about what is the matter that there can be no compromise about a period in bed during the drying out process and about lifelong abstinence thereafter That he will exploit any uncertainties in the mind of the physician goes without saying The earlier he is brought to this realization the less will be the physical domestic and social consequence of his addiction and the permanent damage to his nervous system

CHAPTER XX

POISONING

By Francis E Camps

NOT uncommonly the medical practitioner fails to detect a case of poisoning unless it is brought to his notice by some special circumstance such as a known suicidal tendency or a diagnostic note Such failure can usually be attributed either to an unsuspecting character unwilling to think the worst of human nature or to a clinical training which operates only in terms of naturally occurring diseases when confronted with a problem of differential diagnosis

Poisoning is far from infrequent whether it be suicidal accidental or even homicidal and often the life of the patient can be saved only by prompt diagnosis Broadly speaking there are two types of poisoning—acute and chronic In the former instance there is an immediate urgency whilst in the latter the patient s life can usually be preserved unless terminal symptoms are already present by initial withdrawal of the poisonous sub stance and subsequently by appropriate treatment

The majority of cases of acute poisoning are suicidal The surrounding circumstances should give rise to suspicion although each year deaths occur in which the noxious agent is not only unrecognized but quite unsuspected Occasionally accidental acute cases occur usually from inhalation of carbon monoxide drinking from the wrong bottle (liniments) or access by children (ferrous sulphate) Personal idiosyncrasy to certain drugs is an occasional cause the possibility of which must not be overlooked

Cases of homicidal poisoning are usually unsuspected because the practitioner has subconsciously deceived himself a pheno menon which is well shown by death certificates indicating some natural disease devoid of real clinical basis If the practitioner would admit to himself that on occasions he really does not know

the cause of death and is practising self deception if he issues a death certificate he would avoid inconvenience not only to him self but to others

Chronic poisoning is mostly accidental and includes lead poisoning in its chronic forms

Early diagnosis of poisoning must depend initially upon the use of the senses of sight and smell augmented by common sense and clinical knowledge Thus when called to see a patient who has been unexpectedly taken ill the doctor should always observe the locus from the point of view of possible poisoning such things as empty pill boxes or bottles the smell of coal gas or abnormal vomitus may give the answer After taking immediate measures for resuscitation he should look for evidence of corrosion of the mouth or some diagnostic odour of breath or vomitus

It is obvious too that the clinical condition of the patient may well give some indication should he be in coma (hypnotic) or severe shock (arsenic or phosphorus etc) or sweating (aspirin) At the same time if he is conscious direct questioning may be of assistance although it must always be remembered that in cases of aspirin poisoning for example a person who has taken an overdose may well deny it. As it is axiomatic that *all cases of poisoning are best treated in hospital* immediately resuscitation has been initiated or specific measures have been taken the patient should be removed by ambulance accompanied by a note con taining any relevant information and by material evidence such as empty bottles or containers and samples of excreta such as urine blood or vomit

It cannot be too strongly stressed that unless poisoning is suspected it may well be completely missed and the patient s chances of recovery greatly reduced Early diagnosis may be considered in two stages—suspicion and confirmation

SUSPICION

Suspicion may be aroused by previous knowledge of the patient either on the part of the practitioner or from personal contacts This may take the form of a history of attempted suicide in the past or of recent depression or mental disease This has positive

value only and absence of such information does not of course exclude suicidal poisoning Suspicion may also be aroused by surrounding circumstances such as empty pill boxes or bottles and here again such evidence does not of course exclude coin cidental natural disease

A man was found dead sitting at his desk with a note indicating his intent to commit suicide and a bottle containing cyanide of potassium was in front of him with the cap still screwed up At autopsy no cyanide was found in the stomach contents or blood It would appear that he had died of coronary insufficiency before he could ingest the poison

Nor for that matter does a denial of having taken anything exclude poisoning although a positive admission must be accepted even if there are no apparent ill effects Visual evidence too can be misleading for a person found unconscious in a room with a smell of coal gas may also have taken an overdose of barbiturate The suspicions of relatives must never be treated lightly especially when there is any possibility of a child having had access to drugs or poisonous substances

A practitioner was called to see a child who was alleged to have swallowed a bottle of physeptone prescribed for its father On examination he could observe no untoward symptoms came to the conclusion the story had been exaggerated and recommended expectant treatment He was recalled to find the child in deep coma due to irreversible cerebral anoxia caused by physeptone poisoning from which she later died in hospital

Even when there are no grounds for suspicion in the surround ing circumstances or history there are certain clinical symptoms which must always be viewed with an open mind

Coma (dealt with more fully from the symptomatological point of view in Chapter XVI) always arouses suspicion especially when the patient has been found unconscious in bed or on a chair and when examination reveals no localizing signs Suspicion is deepened if there are or have been hypnotics in the house Signs of **gastro intestinal irritation** such as vomiting or diarrhoea are often significant particularly when they are accompanied by evidence of shock when they arise in the absence of any signs of local abdominal disease when they are limited to one person in a family (arsenic antimony) or when they are accompanied by marked thirst (phosphorus) **Convulsions** may arouse suspicion particularly in children who may have had access to poisonous

plants or berries or when there are known to be anti histamine drugs or belladonna in the house **Sweating** is an important sign especially when preceded or accompanied by vomiting (aspirin) as is **jaundice** without obvious clinical cause (phosphorus) In addition to these symptoms there are certain immediately obvious signs which will be recognized if they are looked for such as corrosion of the mouth (corrosive poisoning) odour of breath (cyanide phosphorus solvents liniment) Once suspicion exists there is no justification for awaiting confirmation before treating the patient unless specific treatment may do him harm should the diagnosis be incorrect For example on one occasion a patient suffering from a cerebro vascular catastrophe was treated with picrotoxin under the mistaken idea that he was poisoned with barbiturates In any event confirmation is usually best carried out in hospital where additional specific and resuscitating measures are available

CONFIRMATION

Confirmation of diagnosis is made by excluding an origin for the patient s symptoms in naturally occurring disease and by clinching the positive diagnosis with specific tests or observa tions Some clinical aspects of differential diagnosis have been dealt with above but a description of the procedure to be adopted in confirming or excluding poisoning may be helpful especially where identification of the poison is essential for proper treatment

In this connection the commonest problem is that of the patient admitted to hospital in coma Admission to hospital is specifi cally mentioned because home conditions are unsuitable for the treatment or the diagnosis of coma caused by poisoning and all cases should be transferred by ambulance as soon as possible after adequate steps have been taken to minimize the risk of irrever sible cerebral anoxia After exclusion of the commoner causes of coma on clinical grounds or if hypnotic poisoning is suspected an invariable routine should be followed

A specimen of urine should first be collected and in addition to routine examination this should always be tested with ferric

chloride solution to exclude the presence of salicylates a further portion being retained for more complex chemical examinations.

A woman attended hospital stating she had taken a large number of aspirin tablets She showed no evidence of any abnormality on clinical examination and was sent home to return if she became worse Some hours later she started to vomit and reported to her doctor The urine now examined showed a strong reaction with ferric chloride and she was sent back to hospital where she subsequently died

A sample of **blood** should be submitted to routine examination and a further portion retained for chemical tests The same considerations apply to a specimen of **cerebro-spinal fluid,** and the **stomach contents** should be examined either in the form of vomit or by means of a sample obtained by suction

Some controversy has arisen as to whether the stomach should be evacuated in patients with barbiturate poisoning in view of the risk of possible inhalation and subsequent pneumonia In the case of barbiturates such as barbitone and phenobarbitone there appears to be little justification for withholding evacuation in view of the fact that the more of the drug that is absorbed the more will have to be excreted With the more rapidly acting preparations which are speedily broken down the need is not so urgent but the presence of a large quantity which may still be absorbed cannot benefit the patient Furthermore the stomach contents may prove to be the most rapid source of identification of any barbiturate present although its presence is readily detectable in the blood or urine provided there are adequate laboratory facilities and especially if an ultra violet spectrometer is available

If the procedure outlined is adopted then the diagnosis of barbiturate poisoning can be established and appropriate treatment instituted Certain other hypnotic drugs however are more difficult to identify and these include carbromal oblivon and the tranquillizers In all cases samples of material should be preserved in sealed and refrigerated containers for later examination

From a clinical point of view it should be possible to diagnose morphine or opium poisoning from the presence of small pupils and slow respiration although it is important to note that barbiturate poisoning may simulate the pupillary appearances

Coma due to carbon monoxide poisoning is usually easy to recognize because of the circumstances in which the patient has been found but in all suspected cases a sample of blood should be collected immediately on admission either into a small bijou bottle (which should be completely filled) or into a vacuum container

Alcoholic poisoning may be suspected from the odour of the breath but should be confirmed by examination of specimens of blood and urine collected at the time

A recent outbreak of methyl alcohol poisoning occurred in New York where a number of citizens celebrating a happy event decided to manufacture a powerful beverage to assist in their festivities To do this a quantity of commercial alcohol was obtained and mixed with eggs and some milk Most of the participants were well after the party but later suffered from vomiting and coma with fairly rapid death Methyl alcohol was recovered post mortem

Symptoms of gastro enteritis if not of bacterial origin may be due to first stage phosphorus poisoning in which case there may be a history of burning sensation in the oesophagus or stomach shock intense thirst and eructation of gas with a smell of garlic Examination of the vomit in the dark should show luminesence and samples of faeces and urine must be collected for chemical examination The second stage of phosphorus poisoning is one of liver failure with jaundice Common sources of phosphorus are pesticides containing elemental phosphorus or zinc phosphide Somewhat similar symptoms may occur from poisoning by fungi (Amanita muscarea Amanita phalloides)

Acute poisoning with arsenic also presents with vomiting abdominal pain and diarrhoea usually with blood in the faeces while shock and collapse may be marked features The commonest source of such poisoning under normal conditions is weed killer In children the accidental ingestion of ferrous sulphate (Fersolate) gives rise to gastro intestinal symptoms with blood in the vomit and faeces followed within twenty four to forty eight hours by signs of liver damage

Convulsions in children may be due to the taking of anti histamine drugs and unless the history lends a clue the identification of this origin for such a non specific symptom may be extremely difficult Except from therapeutic sources strychnine

poisoning is most unlikely unless there is access to agricultural supplies

Sweating is a valuable sign in the diagnosis of aspirin poisoning which is both common particularly because the sale of the drug is uncontrolled and frequently missed Often patients who have taken overdoses not only fail to offer information but categorically deny it when questioned The association of severe sweating with characteristic respiration should always lead to an examination of the urine by the ferric chloride test which should indeed be an invariable routine in cases of coma

CHRONIC POISONING

Most cases of chronic poisoning are due to industrial hazards (see Chapter XXV) but occasionally accidental chronic poisoning may occur from domestic sources The commonest forms are those due to lead and in this connection there is on record one case which occurred in an actress from greasepaint Other examples have been associated with piped water and sucked toys or paint whilst crayons have been the source of arsenic copper and lead In babies chronic poisoning is alleged to have occurred from boric acid in dusting powder and from aniline in marking ink on napkins

The diagnosis of such forms of poisoning is usually made either in the course of clinical examination or as a result of pathological investigations

HOMICIDAL POISONING

It would be remiss not to mention one aspect of homicidal poisoning namely that certain features of illness must always lead to such a suspicion The most significant of these is the history of a patient who is repeatedly ill at home and recovers when he is away or in hospital The possibility should always be given consideration in any illness which shows puzzling features incompatible with any familiar syndrome and it should be stressed that because the patient is expected to die deterioration in his condition is not necessarily due to the primary disease

from which he is suffering This consideration applies particularly where there is an evident change in symptomatology

It cannot be too strongly stressed that nearly every case of homicidal poisoning by metallic arsenic phosphorus or even mercury has not only been undetected in life but unsuspected This is in spite of the cases being under the care of very shrewd general practitioners

SYMPTOMS OF COMMON POISONS

The following is a simple abbreviated list of the common symptoms of frequently encountered poisons together with an indication of what to collect if the practitioner is suspicious It is not suggested that this is by any means comprehensive but that it is merely an indication of a line of thought an attitude which still allows for suspicion

Early Diagnosis of Poisoning

Some Common Poisonous Substances

Poison	*Early symptoms*	*Specimens to be collected for confirmation*
Baby contacts		
Aniline (napkins crayons)	Cyanosis Drowsiness Methaemoglobin	Blood Stomach contents
Boric acid (powder)	Epigastric pain Vomiting Diarrhoea Headache Shock Coma	Blood Vomit Urine
Corrosive poisons Nitric acid Sulphuric acid Hydrochloric acid Oxalic acid	Corrosion of mouth tongue and upper air passages Corrosion of stomach Shock Perforation	Stomach contents
Metallic poisons Arsenic Antimony	Metallic taste Vomiting Abdominal pain Diarrhoea Thirst Shock	Blood Hair Nail clippings Faeces
Aspirin	Fast deep laboured respiration Sweating Drowsiness (not always present until late stages)	Urine (test with ferric chloride)

Poison	*Early symptoms*	*Specimens to be collected for confirmation*
Atropine	Dry mouth Dilated pupils Delirium and hallucinations	Blood Urine Stomach contents
Barbiturates	Coma of varying depth	Blood Urine
Codeine *Heroin* *Morphine* *Pethidine* *Physeptone* and allied synthetic substances	Contracted pupils Slow respiration Coma	Blood Urine Stomach contents
Hydrocyanic acid *Cyanides*	*Smell of breath (bitter almonds) and vomit Anoxic picture—sometimes pink *N B *A certain number of people cannot smell this*	Blood Stomach contents
Cocaine	Exhilaration Incoherence Hallucinations Dryness of mouth Dilated pupils	Blood Urine
Fersolate (Ferrous sulphate)	Irritation of gastrointestinal membranes with blood and mucus in stools Liver damage later	Blood Stomach
Methyl alcohol *Methyl bromide* *Methyl iodide*	Giddiness Sleepiness Pallor Intoxication Slurred speech Visual upset	Spinal fluid
Mercury	Metallic taste Burning feeling in oesophagus and stomach Thirst Abdominal pain Vomiting Disorientation Tremors	Blood Urine Stomach contents
Nicotine	Dizziness Nausea Vomiting Burning sensation in throat	Blood Stomach contents (brown)
Disinfectant Phenol (active agent)	Vomiting Numbness Delirium Loss of memory Corrosion of mucous membranes Dark coloured urine	Blood Urine

Poison	*Early symptoms*	*Specimens to be collected for confirmation*
Phosphates (organic) HETP TEPP Parathion OMPA.	Headache Sweating Salivation Oppression in chest Cramps (abdominal) Convulsions	Blood (cholinesterase estimation)
Lead	Abdominal pain Headache Wrist drop Constipation	Blood Urine Faeces
Phosphorus	Burning feeling in oesophagus	Vomit Faeces
Zinc phosphide	Gastric pain Vomiting Diarrhoea Thirst + + Shock	Urine
Potassium chlorate	Vomiting Abdominal pain Diarrhoea Dyspnoea Cyanosis	Blood Urine Vomit
Quinine	Vomiting Abdominal pain Diarrhoea Ringing in ears Dizziness Dilated pupils	Stomach washings Blood Urine
Strychnine	Twitching Convulsions Relaxation between spasms Fairly rapid death	Stomach washings or vomit Blood Urine
Thallium	Vomiting Pain in limbs at first Later (12 hours) Nausea Vomiting Abdominal pain Some tenderness Ataxia Twitching of muscles	Vomit Blood Urine

Summary

Briefly the steps which lead to the early diagnosis of poisoning may be summarized as follows The clinician must always keep an open mind and be capable of appreciating any unusual feature of the case at an early stage He must make an immediate assessment of the surrounding circumstances in all cases of sudden or unexpected illness The first aim of a full clinical examination is to exclude an origin for the patient s symptoms in

urally occurring disease the second to search for confirmatory ıtive evidence of poisoning In a case where this is suspected t aid treatment must be followed immediately by hospitaliza ı and the patient must be accompanied to hospital by any :vant material or other suggestive evidence Definitive diag ıs will depend on chemical confirmation and for purposes of lence the retention of part of any material examined is clearly :rucial importance

CHAPTER XXI

MALIGNANT DISEASES OF THE FEMALE GENERATIVE TRACT

By Frank Stabler

THE natural history of malignant disease of any part of the body is so variable and so mysterious that it is very easy to draw wrong conclusions either from one man's own observations or from the massive compilation of statistics from many men's experience That patients with clinically more advanced growths do less well under treatment than those with less advanced growths may not be an indication that the former have been growing for a longer time There are some cancers that appear to explode in the tissues and others that seem to creep sluggishly through them Moreover even this characteristic whether it be dependent on the inherent nature of the growth or on the resistance of the host does not give us any indication whether the response to treatment will be good or bad The question then arises—is it really as important to make an early diagnosis as has been supposed? Does delay really affect the prognosis if this depends more on the nature of the growth and of the human being in whom it has arisen than upon the duration of the symptoms? There can be no doubt that the duration of symptoms may bear little relationship to the extent of the growth Three examples serve to illustrate these points

A woman came to the writer in 1938 with a carcinoma of cervix which was estimated as being in stage II (cervix and adjacent tissues) She refused treatment and was not seen again until after the war In 1946 she re appeared and did not at first reveal that she had been seen before Her growth was now estimated as being stage III She responded well to radium treatment and is still well

A woman with no other symptoms than prolapse had a Manchester repair operation The amputated cervix was sent for

routine microsection The pathologist chanced to make his section through a piece of cervix showing a microscopic squamous carcinoma Full radium and x ray therapy was given but she died from recurrence six months later

A woman with stage III carcinoma of cervix in 1934 had a much reduced dose of radium because it was felt that the case was hopeless there being only a paper thin septum between bladder and vagina with massive invasion of the paracervical tissues In 1951 seventeen years later the writer was asked by another hospital for details of her previous condition and treat ment She was in hospital for an attack of bronchitis and there was no sign of growth anywhere

Experiences such as these make one wonder if early diagnosis is worth while The answer is that these are very unusual experiences and one or two such cases should not lead us to adopt a fatalistic attitude With a few exceptions the earlier the diagnosis the better the outlook All malignant growths must originate at a focal point in the beginning and whereas we are powerless to stop the process in some patients there is no doubt that in the majority early diagnosis will lead to more efficient treatment and to a greater proportion of cures Some women will disappoint but others will reward and for the sake of the latter it is necessary for all doctors to devote themselves to an exacting unrelenting discipline to ensure that no patient slips through the net It is no excuse to say that delays are often the fault of the patient—they are but that does not release the doctor from his responsibility to find the lady at the earliest possible moment The ability to perform a repetitive task without error and to continue doing so is as much a test of a man s intelligence as any of the carefully devised conundrums that are now so popular Patients have a right to expect intelligence from their doctors

All diagnosis rests on symptoms and the ability to extract a clear history is essential to a good doctor The first need is to allow the patient to talk It is quite revealing how a garrulous exaggerating woman given her head will expose important symptoms and the experienced interlocutor will know just when to give the touch of the reins that will lead to still further revela tions Finally the reins are taken in a firm hand leading

questions are asked and an answer insisted upon Examination must follow All successful quack doctors listen to the patient s story and make an impressive examination A good doctor need only follow suit to satisfy himself and his conscience

CARCINOMA OF THE VULVA

It is quite surprising in this condition to find at what a late stage patients first report to their doctor A woman will come with a history of a fortnight s discharge and one finds a fungating growth the size of an egg or larger Only vigorous questioning will make her admit that there had been something unusual before this time This is partly due to modesty and shame Many of the sufferers are old women and old women often show a much greater repugnance to the investigation of gynaecological disorders than their younger sisters But there is another reason inherent in the nature of the growth In some cases the growth does burst out very rapidly In the early stages there is nothing more than a trifling enlargement of one labium—no pain no discharge If she reports when discharge first appears there may be a sore less than a centimeter in diameter A week later there is a florid fungating ulcerated mass Once the epithelial surface is broken the growth becomes oedematous and infected so that one can hardly believe the woman s statement of the shortness of the history when one sees a foul smelling cauliflower of growth with the surrounding skin purple and ready to give way Such cases are not difficult to diagnose and could be missed only by gross inattention There are pitfalls Ignorance and modesty often combine to cloud the woman s symptoms By the time such a case reaches the consultant the diagnosis is usually made and the woman can easily be led to give a true history of her complaint, but in the first instance she may give quite a fallacious history She may say she has falling of the womb she has piles she has abdominal pain difficulty in passing water and so on Any old woman starting to make such complaints must be examined It has been taught for so long that epithelioma can arise in a vulva showing leukoplakia that it is sometimes assumed that it only arises in this condition but this is not true Most vulval cancers originate in previously normal vulvae

and a prolonged history of itching is not a necessary concomitant However by the time the patient seeks aid pruritus is nearly always present and is the commonest symptom Although it has been mentioned that the patient is usually old (the greatest incidence is between the ages of sixty and seventy) it should be remembered that one fifth of sufferers will be under fifty Early diagnosis in these atypical cases depends then on paying attention and examining every woman who complains of pruritus and remembering that in the first instance the complaint put forward by the woman may be fallacious

In the typical case diagnosis is easy but not all are typical In the majority the growth begins closely adjacent to the clitoris and as one approaches the perineum cancer is less likely The ulcer with its everted edges may be hidden in the groove between the labia The growth sometimes does not ulcerate and may even be pedunculated when it will look like a skin fibroma Basal cell carcinoma (rodent ulcer) is often of this type There may be a swelling very similar to a Bartholin cyst—a rare lesion in old women

Bowen s disease of the vulva is a rare condition of intra epidermal carcinoma The cells are cancerous but have not yet burst through the basement membrane Clinically it presents as a flat red ulcer with its edges little if at all raised above the surrounding skin

The name Paget s disease of the vulva is applied to another rare condition in which malignant cells grow up from below to invade the skin—the malignancy seems to start in apocrine or sweat glands and advance up their ducts The appearances are of a non ulcerated patch looking remarkably like the ordinary strawberry naevus

The most promising line for early elimination of carcinoma of the vulva lies in the early treatment of certain cases of leuko plakic vulvitis I use the expression certain cases because leukoplakia is not a clearly defined entity The word means white patches and these can be produced by many lesions other than *that which may go on to epithelioma* Moreover the pathologist is of no help in deciding from a biopsy specimen unless growth has already started Almost any chronic irritation of the vulva will produce thickening of the keratin layer elongation of the

rete pegs and hyperaemia of the dermis that will lead to a report of leukoplakia The precancerous condition is diagnosed clini cally Trichomonas irritation will show red soreness of the vulva most marked where the discharge runs over the anterior margin of the perineum Diabetic vulvitis also shows a bright red vulva and tinea will show a red area with scaling running up into the groins Leucoderma is very common there will be symptomless geographic patches of white skin extending out on to the thighs

Another condition which may lead to difficulty is kraurosis vulvae or senile vulvo vaginitis (many of the sufferers are far from senile) In this condition all the changes are those of atrophy The labia minora become small and thin until even tually they cease to exist The labia majora also atrophy Most important is the fact that there are marked atrophic changes in the vagina The rugae disappear the vaginal covering becomes smooth and shining and red atrophic spots appear like petechial haemorrhages around the introitus and urethral orifice The patient usually complains of pain not of itching Malignant change need not be feared

The most common condition mistaken for leukoplakia however is that produced by scratching and rubbing Frustrated and *neurotic women take to this habit until it becomes an obsession* The story is usually of irritation gradually increasing in intensity and at first only at night. The scratching may go on for years until the patient will say she gets little sleep and when she does get to sleep she awakens to find herself scratching She may even have tied her hands to the bed head or worn thick stockings over the hands to stop herself scratching On examination there will be a *uniform* white area of skin centred around the clitoris and in a right handed woman *more extensive to the right of the* organ There may be scratch marks but there will not be any areas of thin atrophied skin—all changes are those of hyper trophy She will submit to examination without a murmur even though the vulva looks very sore This condition is not pre cancerous

What then are the characteristics of the variety of leukoplakia which is a precursor of cancer? The woman will complain of soreness and irritation and though these may be worse at night they are also troublesome when she is busy and active Scratch

ing is limited by the soreness of the skin On examination the patient will be timid and flinching for she knows how easily she will be hurt The process starts inside the labia majora and involves the labia minora and clitoris It will spread out from this on to the perineum and in the anal cleft but will never spread into the vagina or on to the vestibule The menacing feature will be the irregular mixture of appearances presented There will be irregular patches of thick white hyperkeratinizing skin and amongst them thin red shining areas Both the white patches and the atrophied areas will show loss of elasticity to the touch One feels that to pull on this skin would make it crack and indeed there often are cracks The anterior margin of the perineum often shows one of these keens surrounded by heaped up cornified layers This condition and this alone is the one liable to progress to epithelioma So much is this so that it justifies excision of the vulval skin

CARCINOMA OF THE CERVIX

It is a deplorable fact that on the average six months elapse between the onset of symptoms in this condition and the beginning of treatment About half of this delay is due to failure on the patient s part to seek advice In three quarters of the remainder it is due to failure on the part of the family doctor to examine or refer his patient for treatment when significant symptoms are reported The rest (one eighth) is due to mis diagnosis or administrative delay on the part of hospitals *Irregular bleeding* is the first evidence in nearly all cases The reasons why women do not report this symptom are many and varied First women regard the discharge of blood from the vagina as a natural phenomenon and it should be kept in mind that normal menstruation itself is not the exactly regular pheno menon that text books and women themselves would have us believe A group of medical and nursing women who declared themselves absolutely regular and who kept a record for a year of the onset of their catamenia showed a scatter of up to a week on either side of their previously stated interval When it is considered that women accept as normal the likelihood of irregularity of menstruation over the age of forty it is not

surprising that they often delay reporting irregular bleeding A common expression amongst hospital out patients gives an in sight into the attitude Asked about the regularity of their menstrual losses they may reply Oh yes I am regular in fact too regular By this they mean that they are having losses at more frequent intervals than every twenty eight days The only form of irregularity that disturbs their equanimity is amenorrhoea Another cause for delay is that they sometimes think that they cannot come for an investigation of their trouble whilst men struation is going on Yet others have a fear of what the bleeding might signify and avoid the doctor because he might confirm the fear

Delay due to the doctor is less to be forgiven In spite of all that has been said and written about the foreboding of irregular bleeding many women are not examined or referred for months after the date of their first complaint The reasons are difficult to elucidate but some of them become clear after long experience It is curious how some doctors allow their patients to conduct their post graduate education Even though women are prone to ascribe a whole series of symptoms to their age once they pass the forty mark there is no justification for their doctors to do the same Perhaps it is an easy way of cutting short a consulta tion but the danger of such a course is not only that it may lull the doctor into perilous inaction but also that it helps to promul gate the public view that anything and everything may be ascribed to this cause The patient of forty with symptoms of any sort may say to her doctor Do you think it is the change? It will save the doctor a lot of explanation and examination if he agrees with her and from then on each of them is all too often content to ascribe any abnormality to this event The word menopause means a pause or cessation of menstruation and anything other than this is not normal One of the most pernicious and slip shod expressions used by medical men is menopausal bleeding This is a contradiction There could be no such condition Any unusual bleeding in a woman over forty needs investigation and explanation It may be fibroids polypi metropathia but it is not the menopause How much more do these remarks apply when a woman is bleeding after the menopause Medical men must guard themselves against adopting the slip shod modes of

expression that their patients use A woman of fifty five may say I have never had the change—still quite regular By quite regular she means that the losses she is having never occur more frequently than at intervals of four to five weeks but close enquiry may elicit that they are in fact quite irregular

Carcinoma of Cervix in Pregnancy

This deserves special mention The association will be found once in 2 000 pregnancies A maternity hospital with eighty beds is likely to meet one case per year A practitioner attending fifty confinements a year will encounter a case in a lifetime In spite of the attention given to pregnant women nowadays delay is still the rule In early pregnancy the bleeding is attributed to threatened abortion Nearer term it is attributed to placental separation and after delivery to subinvolution The patient is put to bed and the bleeding ceases only to recur when she becomes active again In all cases of bleeding in and after pregnancy an examination must be done to search for a cause In early pregnancy a gentle examination will not make a woman abort if she were not about to do so in any case In later pregnancy she must be examined though that examination must be in hospital in case the source of the bleeding is placenta praevia—but she must be examined After delivery any bleeding other than the diminishing coloured loss that continues for six weeks demands examination

Possible Methods of Earlier Diagnosis

So far attention has been directed to the importance of the doctor paying attention and taking active steps at any time in the presence of bleeding other than normal menstruation Is there any means of influencing the patients themselves to report sooner since the greatest amount of delay is due to them? From time to time it is suggested that education of the public in the early signs would reduce the interval Public lectures and articles in women s magazines and newspapers are recommended to make women aware of the seriousness of irregular bleeding Publicity such as this would do much more harm than good It would give rise to more cancerphobia than there is at present

and it would make it less likely that a doctor would examine his patient if he found himself faced with a hundred frightened women amongst whom there might be one who needed investigation The urge for propaganda is based on a fallacy Logical reasonable men say that if only women knew the dreaded symptoms they would proffer themselves for examination forthwith The fallacy is that most women do not apply either logic or reason when they are faced with the unpleasant situation The natural unreasonable reaction is to think It cannot happen to me or If I go to my doctor he might say it is cancer if I wait it will take up and so like an ostrich the woman will not allow herself consciously to face the possibility that she might have anything so unpleasant That this is true is shown by the fact that the delay in reporting is exactly the same amongst nurses and women doctors as amongst the general public One can only conclude that if the general public knew as clearly as women doctors what the symptoms were they still would not report any sooner

In its earliest stages carcinoma of the cervix does not give rise to any symptoms at all Bleeding is not a sign that a woman has carcinoma of the cervix It is a sign that the carcinoma has broken down from infection necrosis or trauma We should get very poor results in breast carcinoma if we took as the earliest sign the formation of an ulcer or the presence of a discharge Is there no way of making a diagnosis before the growth has reached this stage? Routine clinical examination has been advocated It would not be reasonable to apply this to the cervix only We would need also to search for carcinoma of the digestive tract breast lung and bladder to name only the common ones When one considers the procedures necessary to investigate these systems adequately at intervals of at the most six months it is probable that one would prefer death from cancer To apply it only to those who were rich and excessively concerned with their health might just be feasible To apply it to the whole population would be impossible

There is one method of selecting patients worthy of further investigation in cancer of the cervix and body of the uterus It depends on the fact that malignant cells may be desquamated and may be recognized by trained observers using special stains A

rubbing of cervical cells or a drop of vaginal contents is stained with Papanicolou stain and examined microscopically But once more practical considerations make the investigation of doubtful value under ordinary circumstances A recent trial in Birmingham gave an unbiased picture of the effort necessary and the return to be expected The cytologist needs at least six months special training To search a slide takes about ten minutes and eighteen slides a day is about as much as a man can undertake In 2 250 women one invasive carcinoma was discovered that would not have been found by routine methods. In ten per cent of malignant cases a negative smear was reported However eight patients with carcinoma in situ were found by cytology In this condition there are cells having the appearance of malignancy but still restricted within the epithelial layer and limited by the basement membrane As yet we do not know how many of these pre invasive cancers will proceed to invasion but this is certainly not true of all of them It is evident that a vast amount of effort is needed to discover a small number of cases Perhaps by restricting the type of patient from whom smears are taken less effort would be expended for a greater return One authority has said that the women most likely to develop the disease belong to the lower social groups they tend to marry and to start child bearing when comparatively young and they often have large families It would seem that his first condition gives rise to the others and his others give rise to the first

It seems unlikely that smear examination will lead to any significant alteration in mortality rates This is not the same as saying that it is not worth doing Most centres now take a smear from all women over thirty five years of age attending gynaeco logical out patient clinics Taking the smears is not then a burdensome undertaking for patients or gynaecologists In parts of the world where a doctor may be remote from diagnostic aids a smear may be sent and on the basis of a report those patients can be selected who should be brought into the centre for further investigation

Using a magnifying colposcope it is said that leukoplakia of the cervix and carcinoma in situ can be recognized and the most likely place for biopsy located but it can be said of colposcopy and office biopsy as of cytology that it is unlikely to promote

any great advance in early treatment We return then to reliance on the family doctor to keep alert and to see at any rate that no blame can be laid at his door for delay If public propaganda is useless in bringing the patient to the doctor earlier is there no means of achieving this end? The only hope lies once more in the family doctor He should continue to regard himself not as a mere practitioner of medicine but in truth as a family doctor On him devolves the necessity of educating his flock This he should do not by lecturing or frightening his patients but by showing himself always ready to listen to their complaints and to examine them By doing so he will train them not to fear coming to him with what might appear trivial disorders If he has trained them when they were young women to come to him as soon as they missed a period they will the more readily come to him later in life when they have a period too many

Findings on Examination

In most cases recognition that the cervix is affected by carcinoma is easy The fungating friable growth that bleeds on examination does not give rise to doubt but there are two varieties that are readily missed One is the fibrous scirrhous type of growth In it there may not appear to be any ulceration but *the vagina gives the impression of being shaped like a bell tent* or of not having any fornices It is most likely to be found in old women The other is the endocervical growth Here the cervix feels barrelled and no more exact description of it than this is needed *One of the biggest traps is the impression that* carcinoma of the cervix will feel hard to touch It seldom does and a hard object at the vault of the vagina is more likely to be either the normal cervix or a protruding fibromyomatous polyp *The most characteristic sensation imparted on touching* a carcinoma of the cervix is that it is friable—that one does or could with little force break into the tissue Speculum examination is of little help if bleeding is present and even if it is not *the feel of a carcinoma is more likely to lead to diagnosis* than the appearance Any feeling of thickening of the tissues around the cervix (best elicited by combined rectal and vaginal palpation) is highly suggestive The most characteristic feature of all is that

one s examination induces fresh bleeding, and this sign demands investigation in hospital without delay

A point worth bearing in mind is the possible combination of two pathological conditions Uterine fibroids are so common that they may well accompany carcinoma If as Novak states one fifth of women over the age of thirty possess one or more fibroids certainly one fifth of women developing cancer will have fibroids We must be careful in attributing menstrual irregularities to the benign tumour when this may be only a coincidental finding In emphasis of this warning one can say that if symptoms arise after the menopause and fibroids are found the symptoms should not be attributed to the fibroids until the more serious lesion has been excluded Particularly is this so if the symptom is bleeding *Bleeding after the menopause means cancer until microsection has proved otherwise*

CARCINOMA OF THE UTERINE BODY

Much of what has been written about carcinoma of the cervix applies also to carcinoma of the corpus uteri A curious and quite unexplained change has taken place in the last thirty years in the incidence of cancer of the uterus The proportion of corporeal growths to cervical growths was once one to ten In the year 1919 it was stated that carcinoma of the cervix accounted for over ninety per cent of all cases of malignant disease of the female genitalia In Newcastle in the past three years of 177 uterine cancers forty four were corporeal so that cervical carcinoma was only three times as frequent as corporeal The incidence is greatest in women of fifty five to sixty years of age but it can be met in those in the early forties Once more irregular bleeding should be accepted as of startling importance especially after the menopause The bleeding is often less in amount than occurs from the cervix It is often preceded for some months by a white mucoid or yellow watery discharge This is often unnoticed by a woman still menstruating but after the menopause a woman with such a discharge needs diagnostic curettage unless the discharge is clearly due to vaginitis Clinical examination usually reveals nothing of moment and this fact needs emphasis It is only very late and often not then that

the uterus begins to enlarge There is only one way to find out and that is by exploratory curettage Even this may fail to reveal the growth *and if curettage does not reveal pathology and* bleeding recurs in a woman past the menopause she should be treated as though she had cancer and hysterectomy should be carried out

Unlike carcinoma of the cervix corporeal carcinoma affects parous and nulliparous women impartially and there are some pointers to the type of woman likely to develop the disease It is commoner amongst the obese those whose menopause is late and those who have a diabetic tendency If the menopause has been preceded by endometrial hyperplasia or frank metro pathia carcinoma is rather more liable to occur later suggesting that prolonged imbalanced action of oestrogens may be an aetio logical factor Indeed one fifth of women with an ovarian granulosa cell tumour will develop uterine cancer This should be a warning to those who freely prescribe oestrogenic hormones over prolonged periods

The same caution about the possibility of coincident disease applies as in cancer of the cervix It is very unlikely that a woman with fibroids will bleed from those fibroids after the menopause She is much more likely to have cancer

Similarly a cervical polypus even a simple mucous polypus may have appeared at the cervix because the woman has carci noma higher up in the uterus This is one reason why all women with cervical polypi must be curetted as well as having the polypus removed

OVARIAN CARCINOMA

Delay in diagnosis of this condition is largely due to the fact *that there is a common misconception that swelling of or in* the abdomen is an early symptom It is not By the time a malignant ovarian growth produces a complaint of swelling of the abdomen either there is ascites or the growth is very large Women do not notice swelling of the abdomen or if they do they attribute it to other causes The earliest symptom is vague and persistent indigestion —discomfort around the abdomen a disinclination to eat an awareness that peristalsis is taking place (not amounting to colic) an alteration in bowel habit such as a small and un

satisfactory motion with a feeling that defaecation has not been completed A middle aged woman with symptoms of this nature requires examination She may have carcinoma of the stomach or bowel and she may have carcinoma of the ovary Examination should therefore always include pelvic examination A recent case illustrates these points A woman of sixty two complained of backache and abdominal discomfort Gall stones were suspected but a cholecystogram was normal Gastric achlorhydria was found and a barium meal examination was done Nothing abnormal was found except arthritis of the spinal joints What woman of sixty two will not have arthritis of the spine? Butazolidin did not improve the symptoms and she was referred to a spa for physiotherapy and hydrotherapy As this did not relieve her a special corset was provided When she was seen three years after the onset of symptoms the diagnosis was only too obvious There was a hopelessly advanced malignant ovarian tumour reaching to the epigastrium with ascites

Finally whenever a swelling of any sort is found in the abdominal cavity percussion should be used to determine whether there is any ascites The examination takes only a few seconds but it may indicate the need for urgency in a woman previously thought to have a fibroid or other simple tumour All ovarian tumours should be regarded as matters of urgency for until they have been sectioned there may be no means of determining that they are malignant—and a quarter of all ovarian tumours are malignant The incidence of malignancy is one tenth even in young women and in women over forty rises almost to half

RARITIES

Melanoma of the vulva is not always obviously pigmented but any ulcerated lesion of the vulva is suspect

Sarcoma of the genital organs is almost 100 per cent fatal except where it arises in a fibroid So far as early diagnosis is concerned the only lesson is that a fibroid that grows quickly or one which is painful is not a simple uncomplicated fibromyoma

Chorionepithelioma may follow abortion hydatidiform mole or an apparently normal pregnancy The traps that lead to delay

in recognition of this tumour are first of all the fact that the uterus even in hydatidiform mole is not always enlarged in excess *of the supposed stage of pregnancy in a fifth of the cases it will* be smaller Secondly the biological tests (Aschheim Zondek Hogben etc) are not certain evidences that the growth is present or that it is absent In a recent case with large secondary deposits in the lungs the biological tests were persistently negative All *patients who have passed hydatidiform material should be cur* etted and the scrapings examined within six weeks whatever the tests show

Carcinoma of the fallopian tube is very rare indeed but it has a curiously consistent early symptom—the passage of brown or *slightly blood stained watery discharge in middle aged women*

Conclusion

Throughout this account the emphasis has been on the history *and ordinary clinical examination The high flown techniques of* research and specialist gynaecologists are excellent attempts to recognise malignant disease earlier than in the past but as yet they have made no significant impression on the mass of material Until some simple and reliable test for the presence of malignancy *is discovered the best hope of improved results is for the family* doctor to apply himself carefully and persistently to ordinary clinical methods

CHAPTER XXII

DEAFNESS

By Francis McGuckin

IMPROVED methods of medical inspection nowadays give the doctor greater opportunity to recognize deafness in school children Yet it remains true that hardness of hearing at nearly every age is first suspected by the patient or the parent by relatives or friends by schoolteacher or employer Severe hearing loss in the very young and early disability in later years may give rise to nothing worse than uncertainty an uncertainty which is eventually brought to the doctor for judgement In the environment of this first consultation the defect may not be obvious or it may be masked and hence readily overlooked A child can give little help since he is seldom able to describe his defect and is probably unaware of it In the average consulting room the adult may not seem to be deaf particularly if the background noise causes the doctor unconsciously to raise his voice Moreover straightforward clinical examination will often be wholly negative and it would be unreasonable to expect the practitioner either to possess or to know how to employ specialized otological machinery which for the present purpose includes the standard tuning fork The C128 tuning fork may give the physician some information about vibration sense but it can tell him nothing useful about the patient s hearing How then robbed of the comfort given by the simplest equipment may the doctor hope to recognize early deafness? The answer is that he may go very near to a diagnosis and all the way to recognition of a disability by listening to the history and observing the patient

A young mother is anxious about her child now eighteen months old because grandmother has said that none of her own children behaved thus at the same age The child does not take notice as he should and so there must be something wrong Even

at this age one can usually distinguish the infant who responds to his mother s call from the child who fails to note a loud noise There may be serious doubt but the family statements must be noted—and presently it may become clear that the simple speech of the baby if there be any does not develop Another mother may say that her infant used to show a rhythmic reaction to music but that it ceased after a brief illness which careful enquiry shows to have been characterized by a few days of vomiting Here we must suspect that the illness was not a gastro enteritis but meningococcal meningitis with bilateral labyrinthine destruction If the baby had any speech it has gone The cardinal point however is that the mother noticed *a change in behaviour* In another instance there may be only a chance observation about crying The third child in a family has a very loud cry having no particular pattern a cry quite different from that of the other children Subsequently this infant does not develop baby talk and we now recognize that the unusual cry arises from inability to hear and therefore to control the noise it makes More often anxiety comes because the slightly older child has no speech and here the parents have been running away from a dark suspicion eventually made alarming by the observations of friends and relatives

These are the stories which up to the age of two and perhaps into the third year should make one think of severe deafness The fact of deafness may be confirmed by the otologist and its degree may be roughly assessed by skilled therapists It is essential however to make at least a provisional diagnosis at the earliest possible moment since the mother must be guided in the care of the child in order to mitigate the handicap until education by trained teachers can begin when the team will become child parent teacher One may justifiably be interested to know whether the cause of deafness is hereditary congenital or acquired whether the mother had rubella during pregnancy or whether the infant had meningitis at eight months or mumps at an unusually early age but what one must do is to listen at length to the story recognize the significant phrases and then pursue the suspicion raised until it is proved or disproved

Outside this age group *severe* deafness is easy enough to recognize but it is clear that in small children even gross defects

may be difficult to detect and hence the need to seize every piece of evidence however slender It may not be easy to distinguish deafness from mental defect and occasionally both are present Mental deficiency is considered elsewhere in this book (see Chapter XVIII) and most of things there described which the deficient child is unable to do are done perfectly well by the child who is nothing more than deaf Nevertheless we must carefully discriminate between the frustrated tempers of the deaf child who cannot communicate and the lack of social development seen where there is defective intellect Any doubt about deafness as opposed to deficiency can in most cases be readily resolved by a few sessions in the presence of a skilled observer Parents are immensely relieved to find that their child is only deaf bad though that may be and not mentally defective

After the age of two years it is usually absence of speech delay in its development or poor articulation which arouse parental concern About half of such children presenting between the ages of two and five suffer from deafness Unless some other disability is present the *lack of speech* should make one suspect something bordering on total absence of hearing a disability which if confirmed will demand special education *Delayed development of speech* may imply a moderate degree of deafness perhaps allowing of education in an ordinary school with the use of a hearing aid *Defective articulation* suggests failure at certain pitches usually high tones because the child cannot hear the upper components of speech and so cannot imitate them—and here partial or complete correction may be provided by skilled speech therapy

The partially deaf group of children includes some cases of **bilateral chronic otitis media.** Chronic otitis media may exist without obvious earache and without discernible otorrhoea but it must be recognized that the young patient may present as a case of deafness and it is the hearing loss (where both ears are involved) which is important The pathology can be left to the expert who may or may not be able to help at this stage beyond planning the right course for the future At this juncture a dogmatic statement may not be out of place *Any child who is noted to be deaf in the ordinary household is suffering from bilateral deafness* and a child who is stone deaf in one ear (*e g*

after mumps) is not noted to be deaf at all Indeed the adult who develops unilateral mumps deafness complete and permanent as it is notes the defect purely by accident since there is no subjective local sense of deafness

Clearly bilateral chronic otitis media because it is a pro gressive destructive disease may cause insidious deafness at all ages and it may do so when neither patient nor parent is aware of otorrhoea With this remark the exceedingly complex problem of chronic otitis must be dismissed from the discussion

From the age of three onwards there is a story which is heard more and more often through the early school years and which may be elicited even in adolescence In the beginning the parents say that John is deaf with every cold possibly for three days perhaps for three weeks or they say that John has been consistently deaf for three months but looking back and being wise after the event that he may have been deaf on and off with colds for the last year or more Quite often the mother will say that she is not sure whether John just takes no notice that sometimes he does sometimes he does not There is no mention of earache or of running ears If the boy is eight or ten he may himself add that the ears crackle a bit or that sometimes his hearing comes back after blowing his nose Such a history is indicative of **negative-pressure deafness** arising from corking of the Eustachian ostia by adenoid tissue or tubal oedema secondary to paranasal sinusitis If the air content of the tympana is not refreshed during the transient opening of the Eustachian tubes with each act of swallowing then the tympanic gas is absorbed and the drum heads are in driven by the greater atmospheric pressure outside It should be noted that the word used is in driven not retracted The normal drum head can work efficiently only if it has an approximately equal air pressure on either side If the pressure outside is greater than that inside then the ear must be deafened to some extent

Adenoid tissue is present and obvious in all normal children up to the age of ten or twelve years It may become oedematous from acute inflammation or it may grow to an undesirable size If the inter tubal adenoid mass is shallow it expands laterally like a concertina and so may cork the tubal ostia without giving any of the so called classical evidence of adenoids On

the other hand if the mass projects forwards it may cause nasal obstruction without producing deafness The child who is full of adenoids shows all these features but it is important to note that negative pressure deafness may arise from lymphoid tissue which produces none of the text book features

The history of repeated episodes of deafness perhaps culminating in a prolonged attack is diagnostic of negative pressure in both ears It may be an over simplification to say that such a defect arises either from adenoid tissue or from ostial oedema secondary to sinusitis but it is a good rule for the practitioner The decision to go further may be left to the otologist who sometimes has to advise an examination of the post nasal space under anaesthesia since if he has failed to see the epipharynx and if he has any commonsense he will under no circumstances push a finger up behind a child s palate without anaesthesia

Intermittent negative pressure deafness does little harm and many children have brief attacks quite often unobserved A prolonged attack of several weeks or more however calls for skilled assessment because cicatricial changes may result in static or progressive deafness and sometimes the end result may be invasion of the tympanum by keratinizing squamous epithelium (cholesteatoma) The otologist like the practitioner is often dependent on the history and his findings may amount to nothing more than an oily slightly pink appearance in the drum heads or nothing at all

Sometimes the diagnosis will be made at school and not in the home The teacher wonders why John has fallen behind when he used to be quite bright and she tries the experiment of putting him in front of the class or it dawns on John himself that he does better at the front and he tells his mother Too often the child is just dismissed as a poor lot and he goes on being so because he does not understand why

Now we must turn to **otosclerosis**, an insidious cause of deafness consequent on gradual fixation of the footplate of the stapes This footplate is the last link in the chain of transmission from drum head to inner ear and it is normally attached to the oval window by the elastic annular ligament so that it has considerable mobility In some people the annular ligament is gradually replaced by spongy bone and a fixation deafness is the result

The disease occurs much more often in females than in males The writer has diagnosed it at the age of nine operated for it at the age of eleven performed bilateral operations in the same case at fourteen and seventeen and recognized it in the late fifties *For the practitioner otosclerosis is just a story* For the expert it is the same story with perhaps a quiet voice and a peaceful face in addition subsequently augmented by certain audiometric findings The drum heads look beautiful the post nasal space is perfectly normal and although the patient may have had several nasal operations she never complained about the nose until she submitted to surgery For our present purpose we must assume that the expert does no more than make a shrewd guess perhaps confirmed when he later locates an immobile stapes Nevertheless the competent otologist may well decide what will be the diagnosis and possibly what he will advise in the first few minutes of the consultation

A young stenographer of eighteen finds that there is some difficulty at work When Mr Smith dictates he mumbles and sometimes Mr Thompson is not too distinct Then one of her friends at the office says something about having your ears examined This patient is disturbed but not convinced and she appeals to her doctor In the background noises of the consultation and with the view of two perfect drum heads he feels able to assure her that she is merely imagining things and that her office friend is most unkind The girl however loses her job and she has already told the tale of progressive otosclerotic deafness—*until proved otherwise and it is not otherwise if her* voice is soft and her face one of almost angelic calm She is shielded by her deafness from the continuous barrage of meaningless sounds around her Nor is the undergraduate of twenty two in any better case when he has difficulty at his lectures which are less of a problem only when he sits in the front row an unhappy position in any event He has no catarrh (that meaningless word) no abnormality in the ears and in a noisy background he seems not to be deaf at all Consider also the case of the shop assistant of thirty who complains that he hears some customers and not others or that sometimes the hearing is good and sometimes not so good that he may hear better when the shop is full and less well when there is only one

customer He is describing paracusis the capacity to hear better in a noise a phenomenon indicative of middle ear deafness and in cases where the awareness of a defect is altogether vague almost diagnostic of otosclerosis

Otosclerosis is not only much more common in women than in men but it may first become obvious after a pregnancy and may possibly deteriorate further with subsequent pregnancies The otologist frequently hears the young woman say that her deafness was first suspected by others and quite often by the sister in the maternity ward who made some remark about a disability the patient had never realized This is diagnostic of otosclerosis and it is not a disease to be arrested or ameliorated by terminating a pregnancy or by advising future sterility It is true that male otosclerotics do not suffer a step down with each pregnancy but they deteriorate just the same

I suggest therefore that the diagnosis of this form of progressive deafness should be based essentially on the story of a patient who shows no positive evidence on any ordinary examination which may be expected of the practitioner Since during the last twenty years certain surgical methods of relieving the deafness have been developed, notably fenestration and mobilization of the stapes it is desirable that the diagnosis be made or suspected early rather than late The pathology in most cases remains limited to the region of the stapedial footplate for a few years sometimes for many years and at this stage the chance of successful treatment is considerable because the inner ear is as yet intact So long as the patient retains a quiet voice and a peaceful face there is a probability of reasonable cochlear function but when the voice develops an edge one must consider the inner ear a little suspect and if the voice has lost its quiet character then there is not much chance of surgical success Once the inner-ear begins to fail we have to consider whether the patient s lot can be improved not by operation but by the use of a hearing aid

So far nothing has been said about tinnitus Most otosclerotics have some tinnitus and a few make it the cardinal complaint because the noises interfere with the hearing Yet the majority of sufferers come because of doubt about the hearing and only disclose tinnitus after enquiry

The insidious nature of otosclerotic deafness is commonly such that the patient is unaware that this month is worse than last that this year is worse than last or even that there has been deterioration over *half* a decade The disability often advances so slowly that the subject gradually creeps into her shell and becomes detached from this world She hides her deafness from herself her family her friends and the concealment becomes so much of a habit that she unconsciously tries to hide it even from the surgeon whom she comes to consult *And every otosclerotic is deafer than she or he suspects*

Industrial noise-deafness is a hazard of many occupations and in men it is a common cause of progressive disability Whatever *may be the origin or type of traumatic noise the cochlea first* begins to suffer at about 4 000 cycle range that is to say just *beyond the speech range Above and below this 4 000 cycle* range the hearing gradually becomes impaired and we can say that at or about 2 000 cycles the capacity to discriminate speech deteriorates Moreover the speed of impairment is most important—if it is slow there is a measure of compensation because the subject has time slowly to alter his cortical word patterns whereas if the loss is rapid he quickly becomes deaf for speech In present circumstances we can do nothing about the noise of jet engines or about the racket of compressed air machinery except to implore the designers to do something about equipment which we know not only *may* cause deafness but *will* certainly cause deafness At present both employers and unions avoid the problem like the plague The doctor can however take care of the individual If a man has been in a noisy industry for thirty years and is starting to doubt his hearing and beginning to develop a loud voice he can be warned that further noise trauma may cripple his discrimination for speech Thereafter the decision is his own But if a young man working a tractor with an upright exhaust is already showing noise deafness at the age of twenty one then he must be told the consequences in no uncertain terms This man is unduly susceptible to noise and there are *many such No doctor can do anything about noise* deafness already established but he can and must recognize the hazard and at least try to protect the unfortunate individual who is less resistant than his fellows A patient who after exposure

to noise finds that tinnitus is more severe and more prolonged than that experienced by his fellows is almost certainly a susceptible individual and he should be advised accordingly

This chapter has unwittingly become something like Shakespeare s seven ages a brief survey of the commonest types of deafness from the cradle onwards As we grow older new standards for normal sight are accepted readily enough but less easily do we accept changing standards for normal hearing and still less easily the need for revised ideas about normal balance A celebrated statesman remarking that the torments of seasickness grow less with advancing years was telling the experienced otologist about senile degeneration of his inner ears years before high tone loss made his deafness obvious to the uninitiated Few people however have the opportunity or the experience to make such observation but since most people grow old most must sooner or later make adjustments for changes in hearing acuity For people over fifty we must be prepared to make some allowance for high tone loss **Presbycusis**, or the deafness of age begins to be clinically a significant problem in the late fifties though it is not common until the next decade The elderly patient comes or is brought to the doctor for several reasons the chief of which are failing perception of speech intolerance of loud sounds tinnitus and sometimes vertigo though this is seldom a lone symptom The gradual failure in speech perception is very often considered by the patient to be the fault of other people—everyone mumbles these days actors cannot declaim as they did his wife will not speak distinctly and so on Scientific observations about the audiometric response are useful enough in their way but once again a steep loss for high tones is not crippling if it has taken ten years to develop but the same finding is important if it has developed quickly In the former case the patient may do quite well for years if his relatives will speak slowly and distinctly or he may respond usefully to a hearing aid In the latter case there will often be intolerance of noise above a certain level because the inner ear *cannot sort out the jumble which reaches it* and in this event a hearing aid will merely be a source of annoyance This dazzling of the cochlea with too much sound is now called recruitment and it is indicative of failure in the end organ of

hearing itself usually a fairly rapid failure in the elderly patient But it is no new discovery A hundred years ago Anthony Trollope told of the old lady whose ears were so tormented by the noise of the Victorian equivalent of a cocktail party that she would have been better at home and in her own bed We can say of this lady that she had senile degeneration of the cochlea and that she would have regarded a hearing aid as an invention of the devil

So long as both labyrinths are failing slowly and equally we may expect only gradual failure to perceive the consonants in speech If both labyrinths take a sudden step-down the patient may complain for a time of severe tinnitus and severe deafness with a semblance of improvement in the hearing when the tinnitus subsides If however one labyrinth fails rather quickly the patient may complain of a series of attacks of vertigo as well as of annoying tinnitus in one ear

In general age brings with it a progressive degeneration of labyrinthine function usually first noted in the cochlea which is concerned with the finer ripples of fluid arising from sound waves and later in the spirit levels of the vestibular segment which respond to the grosser fluid waves consequent on body movement So we arrive at the tottering old man unwilling to go out in the dark because he is left only with his faulty joint sense and his failing eyes the old man with the childish treble of Shakespeare shouting into his own ears because his air conduction is so much less impaired than his bone conduction

CHAPTER XXIII

PROGRESSIVE FAILURE OF VISION

By A G Cross

THE sense of vision is constantly used in the ordinary occupations of life and it might be imagined that any diminution of visual acuity would be immediately obvious to the person in whom it occurs The practice of ophthalmology however disproves this presumption The ability to see is vested in two eyes and gradual deterioration of vision may proceed for a long time and to a severe degree especially in one eye before the person complains of any disability Sudden loss of vision is much more readily appreciated

Vision comprises visual perception of objects which are near or distant in full or diminished illumination and adequate visual sensation also demands a satisfactory field of vision so that when the subject is looking straight ahead he is aware of what is going on at the sides as well as above and below Early visual failure may involve any of these various essential components and careful examination of all is required for the early diagnosis of progressive visual failure The later stages of visual deterioration usually involve more than one of the components of vision though in most cases one is concerned to a greater degree than others and some part may remain unaffected until very late in the condition The awareness of visual failure is less acute in the elderly and also in the mentally defective and complaint of defective vision in such persons may be long delayed

Progressive failure of vision must be regarded as a steady deterioration of the sense of vision due to organic changes in the eyeball or in its nervous pathways Errors of refraction should generally be excluded from consideration since they represent only a relative defect and can easily be overcome by the use of correcting lenses worn in spectacles The causes of progressive visual deterioration are therefore pathological pro

cesses occurring within the eyeball and visual pathway which may or may not be associated with disease in other parts of the body There are many causes but the important ones are cataract glaucoma retinal degenerations (of which the most important are macular degenerations and retinitis pigmentosa) myopia and diabetic retinopathy Less common causes of progressive failure of vision are optic atrophy and uveitis

CATARACT

This condition which quite needlessly produces a great fearfulness in the lay mind is the result of opacification of the lens of the eye which can be due to various causes These are congenital changes trauma opacities in association with other abnormalities of the eyeball such as uveitis and retinitis pigmentosa opacities caused by general diseases such as diabetes and hypoparathyroidism lens changes of toxic type due to naphthalene ergot and other substances or merely senility The last so called senile type is the most common variety of cataract and is responsible for more cases of progressive deterioration of vision than any other cause It usually occurs after the age of sixty years but sometimes appears earlier

The early symptom of cataract is blurring of vision which varies in degree according to whether the opacities are concentrated in the central area or in the periphery of the lens When the opacities are central it may be noticed that the blurring is more marked when the light is bright and the pupil dilated Most patients complain in the early stage of cataract that there is discomfort described as dazzling in bright light This is the result of irregular reflection of light into the eyes from the opaque spots in the lens and some comfort can be achieved by wearing spectacles of a dark tint which not only limit the amount of light passing into the eye but also allow the pupils to dilate to attain the maximum vision Patients with progressive cataract may have a rapidly changing error of refraction especially tending towards greater myopia and a history of frequent alteration of glasses especially of the distance correction in an elderly person suggests changes in the lens which are likely to lead to increased opacification It is noteworthy that the very common

symptom of spots moving about in front of the eyes especially in a bright light is rarely due to cataract. Patients with early cataract usually notice more difficulty with distant vision than with reading and this is only partially due to increasing myopia because it persists when the best spectacle correction is worn.

Cataract can easily be seen when the eye is examined with the pupil dilated by a drop of two per cent homatropine hydro bromide and one per cent cocaine hydrochloride by looking along a beam of light shone into it either by an ophthalmoscope or retinoscopy mirror The cataract is visible as a collection of dark spots or lines against the red background of the fundus or as distortions of the fundus pattern which occur as a result of the refractile changes characteristic of the sclerosis of the lens which is so common in the elderly Examination with the slit lamp and corneal microscope at this stage may reveal characteristic formations and types of opacities which may assist in identifying the cause of the cataract The well known and easily recognized appearance of a completely opaque white lens in the pupil is an advanced stage and characteristic of that state of cataract which is known as maturity The red fundus reflex seen by ophthalmoscopic examination becomes lost as the cataract approaches maturity but it is sometimes surprising how much a patient will continue to perceive through what appears to be an almost opaque lens provided that illumination is good

The diagnosis of cataract is not one which causes difficulty in the majority of patients but it is not always easy to be certain whether the defective vision is due entirely to the cataract or whether some other pathological condition such as glaucoma or macular degeneration may also be present The assessment of any case of cataract is not complete until the cause of the cataract has been determined and progressive cases occurring in younger patients may require extensive investigation to ascertain the aetiology

Some amount of lens opacity is present in almost all patients over the age of sixty years When the opacities are in the periphery little visual embarrassment is noticed and even if a few small opacities trespass on to the pupillary area they cause no discomfort except for the very slight fading of vision which is regarded by most people as inevitable in later life Progression

of the opacities to cause serious visual embarrassment occurs in only a minority of elderly persons

GLAUCOMA

Glaucoma is a condition in which the pressure of the eyeball usually called the intra ocular pressure is raised This pressure is variable in health in different individuals and in the same person at different times of the day A figure above 32 mm Hg is usually regarded as abnormal and a variation of more than 4 mm Hg during the twenty four hours suggests that a patho logical condition is present Accurate measurement of the intra ocular pressure may present difficulties It can be estimated by instruments A cannula in the anterior chamber attached to a manometer is the most accurate method but it is only applicable in the laboratory The Schiotz tonometer gives a reading of the pressure from the amount of indentation of the eyeball which is caused when a known weight is placed upon it The usual method of estimation is by palpating the eyeball with the index fingers of the two hands and assessing the amount of fluctuation the patient looking down to relax the eyelids The usual method does not record a figure for the intra ocular pressure but after adequate practice it enables the clinician to decide whether or not the intra ocular pressure is raised

Glaucoma occurs in a number of different forms Congestive or narrow angle glaucoma causes a visual defect of rapid onset and is usually associated with severe pain secondary glaucoma is the result of some other pathological state of the eyeball such as inflammation of the uveal tract haemorrhage or neoplasm chronic simple glaucoma is a condition of insidious onset which is steadily progressive in the absence of treatment This last condition is the only form of glaucoma which requires considera tion as a cause of progressive visual failure

Chronic simple glaucoma is characterized by a rise of intra ocular pressure which is usually not greatly above the normal and which often varies throughout the twenty four hours This diurnal variation of the intra ocular pressure throughout the day may be greater than the 4 mm Hg mentioned above The rise of intra ocular pressure may be present for a considerable time

without causing any symptoms and during this latent period it may cause atrophy of some of the fibres of the optic nerve resulting in characteristic changes in the field of vision The most usual visual field change is the development of an arcuate scotoma which runs from the blind spot above or below the fixation point to reach the horizontal meridian in the nasal field but peripheral field changes are also found Associated with the optic atrophy and the visual field changes the optic disc may protrude backward to form the characteristic appearance of cupping when the fundus is viewed through the ophthalmoscope This characteristic cupping of chronic simple glaucoma must be distinguished from the physiological cupping which is found in many normal optic discs from the congenital abnormality called coloboma of the optic disc and from the senile cupping which occurs in many elderly people The cupped disc of chronic simple glaucoma is pale in colour and cupping extends to its margin The retinal vessels can be seen on the floor of the cup and then having ascended the wall they can be observed at a different level on the retina Chronic simple glaucoma usually affects people in the second half of life though rarely it may occur at an earlier age It is commonly bilateral the condition of one eye being more advanced than that of the other

Chronic simple glaucoma as has been stated is often symptomless in the early stages and it may reach an advanced state before it is diagnosed This late diagnosis causes difficulties in the provision of efficient treatment and as a result the condition causes many cases of blindness The earliest symptom in many cases is of indefinite visual failure which causes the patient to seek an examination for the provision of new spectacles when the cupping of the optic disc may be discovered at routine ophthalmoscopic examination The typical arcuate scotoma may then be found at perimetric examination Perimetry is not usually undertaken as a routine procedure in all patients but only when it is indicated by some pertinent symptom or by the fundus appearances In most patients chronic simple glaucoma does not affect central vision until a late stage and observation of the optic cup and the visual field changes is essential for the diagnosis Some patients with good powers of observation may notice the arcuate scotoma as a blind spot when they are reading

but this is unusual Other patients may notice difficulty with reading but may not realise the reason and a complaint of difficulty with close work in an elderly patient who can read 6/6 on the Snellen chart and who has recently had new (and presumably correct) glasses should lead to a suspicion of chronic simple glaucoma A history of frequent changes of reading glasses over a period of a few months should also suggest the possibility of early chronic simple glaucoma The insidious pro gression of this disease and the need for early diagnosis is an important reason for advising that all elderly patients should have a regular eye examination at least every two years and more frequently if there is a family history of glaucoma Patients with chronic simple glaucoma require comprehensive investiga tion by an ophthalmic specialist who after the diagnostic fundus examination for cupping of the disc and examination of the visual fields for characteristic changes will undertake other special examinations *Gonioscopy* allows observation of the angle of the anterior chamber where drainage of aqueous occurs by means of the special contact lens and the corneal microscope and is useful in showing whether the angle is wide or narrow and open or closed these facts are necessary in assessing the correct treat ment. *Phasing* which is the two hourly examination of the intra ocular pressure for twenty four hours with Schiotz tonometer is required to decide whether the intra ocular pressure is raised or whether the diurnal variation is unduly large *Tonography* which is the measurement of the speed of outflow of the aqueous humour from the eyeball is also useful in deciding the correct treatment, serious diminution of the rate of outflow being an indication that surgical measures should be considered

Chronic simple glaucoma is a condition which must always be considered in the diagnosis of any patient who complains of difficulty with vision The majority of cases are not recognized until obvious glaucomatous cupping is present in one eye because they have not previously complained of visual dis comfort Unfortunately this indicates a moderately advanced state of the disease and unceasing vigilance is necessary in order to diagnose the condition at the earliest possible moment. Any patient who shows the least suspicion of cupping should undergo

examination of the peripheral and central visual fields and should have full investigation of the intra ocular pressure

RETINAL DEGENERATION

Retinal degeneration is usually local rather than general and commonest at the macular region General retinal degeneration occurs as the so called retinal abiotrophy of which the most common form is retinitis pigmentosa more correctly known as pigmentary degeneration of the retina

Macular degeneration occurs in various forms and at all ages but it is most frequent in later life when it is described as senile macular degeneration It has hereditary tendencies The macular region shows excessive pigmentation atrophy and exudates and in any particular case any one of these three features may pre dominate That type which shows a massive exudate covering the whole macular area is called disciform degeneration of the macula Haemorrhages may be present and they appear to come from the choroidal vessels The cause of macular degeneration is unknown but it appears to be the result of degenerative changes in the blood vessels forming the choriocapillaries of the choroid in the macular area Both eyes are affected but the retrogression of one is usually more advanced than that of the other Macular degeneration is characterized by slow and painless visual deterioration with occasional episodes of more acute failure associated with haemorrhages Reading and close vision are more seriously embarrassed than distant vision Distortion of vision may be noticed in the early stages but this disappears as visual acuity deteriorates In the late stages the patient may become aware of an area of complete central blindness and will say that he can only see things if he doesn t look at them which indicates that he is using paramacular vision The diagnosis of macular degeneration can only be made by ophthalmoscopic examination when the typical appearances at the macula are found This investigation may be difficult because the pupils are often very constricted in elderly patients and lens opacities are frequently present The pupil should be dilated so that careful inspection of the macular area can be undertaken This is particu larly important in cases of incipient cataract because a stage will

be reached when the retina can no longer be seen and when macular degeneration can no longer be diagnosed Removal of a cataract in the presence of macular degeneration may not cause significant improvement of vision and an unnecessary operation may be avoided if the macular lesion has been diagnosed in the early stage of the cataract. There is no treatment which cures or prevents deterioration of vision in these patients but the disease is not one which leads to absolute blindness The visual field is retained and this enables the patient to move about and be independent in his own surroundings Detailed vision however becomes quite impossible

It is characteristic of macular degeneration that it tends to progress to a considerable degree in one eye and to a less extent in the other before any complaint of defective vision is made It appears that the normal vision is more than adequate for the ordinary duties of life and that it is only when some deterioration has occurred that the threshold is reached which provokes complaint

Retinitis pigmentosa (pigmentary degeneration of the retina) is a generalized retinal degeneration which usually begins in the equatorial region of the retina in the early part of life and apparently affects the rod cells before the cone cells It is characterized by the appearance of retinal pigmentation which takes the form of fine lines irregularly joined together—spidery pigmentation Areas of retinal atrophy may be present, but exudates do not occur The optic disc becomes pale in the later stages and the arterioles much attenuated Lens opacities may also occur as a late manifestation The earliest symptom is usually *defective vision under conditions of diminished illumination* A number of patients with retinitis pigmentosa were discovered at the beginning of the Second World War when the blackout was introduced and they were found to be unable to walk about unescorted at night. Other causes of a similar reduction of visual acuity under conditions of poor lighting are vitamin deficiency and psychological disturbances In all cases the degree of defect can be measured by estimating the rate of dark adaptation and by more practical methods such as the Livingston Hexagon which has been employed in the Royal Air Force Retinitis pigmentosa is steadily progressive but central vision in full day

light usually remains normal until the disease has reached an advanced stage The peripheral fields are gradually constricted until only a few degrees of field remain around the central fixation area this is known as tubular vision The diagnosis a suspicion of which should always be raised by the complaint of defective vision in diminished light depends upon ophthalmoscopic examination of the fundi and the observation of the typical pigmentation A similar type of pigmentation may sometimes be seen as a result of *healed choroiditis* but some choroido retinal scarring is usually present in such cases and the arterial attenuation is not so marked The course of retinitis pigmentosa may be very prolonged but extending over a period of twenty or thirty years it inevitably leads to blindness and no treatment has been conclusively proved to delay this final result It is a hereditary condition and is inherited in various ways Whatever the hereditary pattern it necessarily follows that the disease occurs more commonly when there is parenteral consanguinity and this factor should always be considered when marriage is proposed between near relatives

The diagnosis of retinitis pigmentosa is usually made in the teens or in early adult life and it is frequently found at routine examination in the absence of relevant symptoms The presence of symptoms is an indication that the condition has advanced beyond its earliest stages

MYOPIA

Myopia or short sight is a common condition but it is only in exceptional cases that it leads to progressive failure of vision It is usually due to the eyeball being longer than normal so that rays of light which enter the eyeball are brought to a focus in front of the retina The cause is unknown but there is a familial tendency It usually commences in childhood and it increases during the period of growth It rarely becomes worse after the age of twenty years Myopia is usually suspected because teachers find that a child is having difficulty in seeing the blackboard or with older children because of complaints that they cannot see properly They may have difficulty in reading bus numbers or in recognizing people they meet Some patients are observed

to hold their reading matter very close but this may be habitual and unassociated with myopia Examination of the refraction establishes the diagnosis and suitable spectacles usually correct the vision Changes occur in the fundus especially in the higher degrees of myopic error and the defects appear at the posterior pole The first change is the myopic crescent which appears as a white crescent on the outer side of the optic disc This may later extend all round the disc and the macular region may become involved These cases of progressive myopia suffer deterioration of vision which cannot be corrected by glasses and in the worse cases blindness ensues This may be precipitated by macular haemorrhage and by retinal detachment Most myopic individuals require no special management except the routine examination of refraction at regular intervals Those who have an error of ten dioptres of myopia or over should be regarded as liable to visual deterioration and their activities should be controlled In the case of children some thought should be given to their occupation in life It is essential that they should avoid work involving strenuous manual exertion since this predisposes to macular haemorrhage and to retinal detachment Close work on the other hand is not detrimental but patients with high myopia should undertake administrative or industrial work which they could continue should they become blind

DIABETIC RETINOPATHY

Diabetic retinopathy is the most important ocular manifesta tion of diabetes mellitus and may progress to blindness Its onset is related to the duration of the diabetes and it shows specific changes on ophthalmoscopic examination The earliest change is the formation of capillary micro aneurysms which appear to be formed by adhesion of the contiguous walls of capillary loops in the inner nuclear layer of the retina These can be seen ophthalmoscopically as small red dots Haemorrhages may occur in the deeper retinal layers and exudates are usually present in the external nuclear layer These are almost white with clear cut edges and they usually occur at the posterior pole of the eyeball They are discrete at first but may later coalesce Severe haemor rhages may occur at any time and these may invade the vitreous

and give rise to organization (retinitis proliferans) and to retinal detachment The diagnosis of diabetic retinopathy depends upon the ophthalmoscopic appearances of the retina which are quite characteristic and differ in several respects from hypertensive or arteriosclerotic retinopathy A minority of cases of diabetes are diagnosed as the result of an initial observation of diabetic retinopathy but in the majority of cases the condition occurs in established diabetic patients The precise cause of the retinopathy is uncertain and in particular the reason for the rapid deterioration in some patients is quite unknown There is no specific treatment which will influence the condition or which will prevent its progression The control of the diabetic condition by diet and the use of insulin where necessary is essential and many patients with this condition deteriorate so slowly that they complete their lives without the visual acuity being seriously affected It is the minority which deteriorate steadily and in them blindness may ensue at a fairly early age

OPTIC ATROPHY

Atrophy of the optic nerve fibres causes variable pallor of the optic disc due to disappearance of its small blood vessels This atrophy may be primary when it is unassociated with any other preceding or accompanying disc changes and due to damage to some of the nerve fibres between the optic disc and the external geniculate body or it may be secondary when it follows neuritis of the head of the nerve or papilloedema due to raised intracranial pressure The disc in primary atrophy is white with clear cut edges but in secondary cases it has a blurred margin and some organized tissue is present which obscures the structure of the lamina cribrosa Optic atrophy can occur at all ages and from many causes Some of the more common causes are *head injury meningitis optic neuritis retrobulbar neuritis pressure from orbital and cerebral tumours* the effects of certain poisons such as lead tobacco arsenic quinine and salicylic acid the condition of *arachnoiditis* and the after effects of *papilloedema* due to increased intracranial pressure Progressive visual failure in contrast to a sudden deterioration of vision occurs in pressure from tumours from toxic causes when the poison is ingested in

small dosage but over prolonged periods in arachnoiditis and after long continued papilloedema The essential symptom of optic atrophy is visual deterioration, and some patients observe definite defects in their field of vision On examination the pallor of the disc can be observed by ophthalmoscopic examination Difficulty may arise in the earliest stages in the differentiation of mild degrees of atrophy and in such cases comparison of the two discs may be helpful Examination of the visual fields may assist in deciding the nature of the atrophy The diagnosis of optic atrophy is an indication for immediate specialized neuro logical investigation which may involve radiological examinations of various kinds and lumbar puncture in addition to routine clinical examination Atrophy of optic nerve fibres is an irrevers ible process and it is essential that its cause is discovered and treatment instituted before undue damage occurs to the visual acuity Some of the causes of optic atrophy involve a danger to life itself and this renders diagnosis a matter of even greater importance

TOXIC AMBLYOPIA

Various substances can cause progressive visual deterioration if continuously ingested by any route and they may be regarded as exogenous toxins of the optic nerve They include methyl alcohol lead arsenic and ergot but the most common and important of them is *tobacco* Tobacco amblyopia can occur as a result of smoking or chewing tobacco or by inhaling it as a snuff but it is most commonly found in pipe smokers especially when they smoke the stronger forms of tobacco Tobacco amblyopia occurs in persons who have smoked large amounts of tobacco over a long period of time and it is usually taught that nobody develops tobacco amblyopia until he has smoked his own weight of tobacco It is unusual for tobacco to be the only factor in causing amblyopia Most persons who develop this condition have some other pathological condition such as diabetes mellitus or anaemia and in addition to tobacco addiction they may have alcoholic tendencies Tobacco amblyopia is character ized by a steady deterioration of the central vision of both eyes with a selective defect for red and green Some pati nts notice

this defect and complain of difficulty in identifying the colour of traffic lights The diagnosis of tobacco amblyopia is to some extent a matter of exclusion because examination must show that the more common causes of bilateral visual defect in elderly persons such as cataract and macular degeneration are not present These patients show no abnormality of the fundus except for some pallor of the optic disc in the later stages There is a scotoma for red and green involving the whole central area and connected to the blind spot The history of fairly heavy tobacco smoking if available usually leads to the diagnosis which is finally confirmed by at least partial recovery of the visual acuity after the use of tobacco by any route has been stopped for six to eight weeks Return to tobacco causes recurrence of amblyopia and such persons must become life long abstainers

UVEITIS

Uveitis an inflammation of a part or of the whole of the uveal tract (comprising the iris ciliary body and choroid) is one of the commonest eye diseases and it occurs in many and various forms It may have an acute onset with pain and sudden deterioration of vision or it may begin gradually with only the mildest blurring of vision and without pain The important signs of the disease are the presence of keratic precipitates (K P) on the back of the cornea, adhesions of the iris to the anterior lens capsule (posterior synechiae) and vitreous opacities There is also an increased amount of protein content which causes opales cence of the aqueous humour and inflammatory cells are present in the anterior chamber of the eyeball these signs can be seen only by examination with the slit lamp and the corneal micro scope *Keratic precipitates and posterior synechiae* can be seen with an ordinary magnifying glass if the eye is examined with a bright light Uveitis is due to a multitude of causes and a full clinical radiological and pathological examination is necessary to discover the aetiological agent. Amongst possible causes are *tuberculosis sarcoidosis venereal disease intestinal infection focal infection* of any kind or a protozoal agent such as *toxo plasmosis* The acute cases usually clear up rapidly but chronic cases may persist over a prolonged period Such cases may be

complicated by the occurrence of lens opacities macular oedema or even by softening and shrinkage of the eyeball which is known as phthisis bulbi all of which cause progressive deterioration of vision Early diagnosis and energetic treatment are therefore most important in the treatment of uveitis since delay means progression of the inflammation organic damage to the eyeball and irreversible defects of visual acuity

It is apparent that progressive deterioration of vision may be due to many causes The most important aspect of the condition is that its onset is often insidious This is unimportant in conditions like cataract which can be treated by surgical means even in the advanced stages and in macular degeneration where treatment is of no avail but in chronic glaucoma and in optic atrophy in particular the whole success of treatment depends upon early diagnosis and it is essential that all patients complaining of visual defects should be thoroughly investigated to diagnose or to exclude the presence of these progressive diseases

CHAPTER XXIV

SOME COMMON SKIN DISEASES

By S T Anning

Though not usually a life saving matter early diagnosis of the common skin diseases is important in that it may save months or years of discomfort and disability and greatly reduce absence from work The early diagnosis of disorders of the skin should be easier than that of other conditions for we can see and touch the lesions and they readily lend themselves to histo logical or bacteriological examination For this reason there may be a temptation to make a rapid or spot diagnosis Though this may provide a euphonious and important sounding Greek label usually merely descriptive it tells us little about the patient or the cause of his trouble The patient even though he merely has boils or vulgar warts must be treated as an individual with a general medical problem Why for example has his resistance to certain ubiquitous organisms broken down? As in most other branches of medicine *the taking of the history is the most important part of the examination* and in dealing with disorders of the skin one often learns much by asking the patient his own views about the cause of his condition

The skin is not merely an important organ in itself which preserves our internal environment and protects us from that outside controls our body temperature and is one of our chief sensory receptors It is part of the body as a whole and reflects many internal changes—mental biochemical and structural Its examination is therefore often of importance in the early diag nosis of internal disease

Disorders of the skin are of two kinds the first **pathological conditions** such as congenital defects acute subacute and chronic infections and neoplasms benign or malignant and the second **disorders of function or reactions of the skin** The first steps in diagnosis is to decide into which group the condition with wh ch

we are confronted is to be placed Unlike the pathological diseases the disorders of function which are by far the more common are labile varying even from day to day clearing and relapsing *They are an expression of the patient s temperament* and personality the way he has been moulded by heredity and by the life he has led representing the reactions of the skin to external or internal stimuli—an individual affair For example in a group of medical students before an examination some may be found to develop eczema some alopecia areata or psoriasis lichen planus or profuse sweating just as others have diarrhoea migrainous headaches palpitation or dyspepsia *Some individuals* are more sensitive than others in the physiological sense of responding more readily to stimuli whether emotional aesthetic or physical and it is in such subjects that the disorders of function are usually observed In the skin the simplest mani festations of such disorders are erythema (as in blushing and rosacea) and the over production of sweat or sebum

It should never be forgotten that the functional dermatoses may be reactions in the skin to some organic disease If a middle aged or elderly person with no previous history of skin trouble develops such a syndrome a most careful examination must be made to rule out such conditions as hypertension renal failure metabolic disease and new growths

PATHOLOGICAL CONDITIONS

The pathological conditions need not be considered at great length since their early diagnosis is usually straightforward while if difficulty arises histological or bacteriological investigation is likely to elucidate the problem

Of birthmarks the **haemangiomata** are most common Early diagnosis is important only in regard to prognosis If the haeman gioma is of the strawberry mark variety deep red a little raised and rarely more than a centimetre or two in diameter the parents minds can be set at rest for all such naevi disappear *spontaneously within the first seven years of life Treatment is* unnecessary The variety known as a port wine stain is how ever persistent and not very responsive to treatment With port wine stains of the forehead the underlying meninges may

be affected by a similar form of haemangioma sometimes complicated by epileptic convulsions

Impetigo contagiosa is one of the more common acute infections of the skin It may result not only from contact with some affected person but it may be secondary to a discharge of the ear from otitis media or a nasal discharge from chronic infection of an antrum When impetigo occurs on the head pediculosis capitis should always be looked for especially in children of pre school age Early diagnosis may prevent the infection spreading to other members of the family The condition usually starts with the appearance of flat thin walled vesicles first clear then purulent which burst dry up and form yellow crusts They give the appearance of being stuck on to the skin and are surrounded by a little erythema

Erysipelas is another acute infection of the skin which must be mentioned It may recur in the same area and an appreciation of its complications makes it unnecessary to stress the importance of diagnosis With each attack lymphagitis is likely to occur and each may be followed by increasing and permanent obstruction of lymphatic vessels resulting eventually in lymphoedema which may reach the degree known as elephantiasis Erysipelas is an infection of the lymphatic vessels of the skin by *Streptococcus pyogenes* which often gains entry through fissures Thus cracking of the nostrils may lead to erysipelas of the nose and after several attacks to chronic swelling of nose and upper lip A similar state of affairs may affect the ear while with tinea pedis fissuring between the toes may lead to recurrent erysipelas of the foot and leg which may ultimately result in considerable swelling of the whole of the lower limb Such attacks are marked by a high temperature and rigors in addition to the local signs but they are usually easily controlled by injections of penicillin An important aspect is diagnosis of the cause of recurrent attacks for if fissuring is responsible it must be dealt with

Chronic infections such as tuberculosis syphilis and those caused by some fungi often produce a **granuloma**—a purple indurated lumpy lesion which may become ulcerated Granulomata are uncommon and require little further mention here though the bright red rounded pyogenic granuloma (usually about two to five mm diameter) must not be ignored Granulo

mata may also be provoked by iodides and bromides and by certain foreign bodies in the skin such as silica and beryllium as well as by puncture of the skin with the point of an indelible pencil or a ball pointed pen

Ringworm infections sometimes provide a difficult diagnostic problem An eczematous condition (pompholyx) with a bullous and scaling eruption of the feet may be indistinguishable clinically from tinea pedis though if it is unilateral tinea is more probable It is not difficult to examine microscopically scrapings from a bulla or a scaling area after mounting the material on a slide in a twenty per cent solution of potassium hydroxide and warming for ten minutes The discovery of mycelium establishes the diagnosis It is not generally appreciated that tinea pedis especially when caused by *Trichophyton rubrum* introduced during the last decade or so from the Middle and Far East may not only produce blistering and scaling of the feet with dystrophy of the nails but may also be responsible for scaling patches on any part of the body roughness of the palms and ano genital pruritis This latter trouble may also be the result of an infection with *Candida albicans* especially in diabetics

Occupation may provide a pointer to the diagnosis of some acute and chronic **infections of the skin,** for certain infections are associated with particular trades leather workers and anthrax herdsmen and cattle-ringworm fishmongers and erysipeloid shepherds and orf (a virus infection of the mouths of sheep causing a moist grey firm papule usually seen on the hand) Butchers veterinary surgeons and farmers may develop warty tuberculosis of the hands All these conditions though not common are constantly encountered in this country

Benign new growths of the skin will not be considered here but squamous and basal celled carcinoma and malignant melanoma demand our attention **Squamous-celled carcinoma** is more likely to appear in skin which has been long exposed to sunlight or in contact with certain industrial oils or tars and in skin previously damaged by radiotherapy thermal burns or chronic ulceration At first the tumour is warty tending to bleed readily and later it becomes ulcerated Lymph nodes may become involved In the differential diagnosis **molluscum sebaceum** or kerato acanthoma must be mentioned Until a few

years ago this condition was thought to be an acute epithelioma of low grade malignancy It occurs on exposed skin commonly on the face and often at the site of slight injury (the writer recently saw one on the nose following a peck by a budgerigar) Rapidly increasing in size over a period of a month or two it becomes a dome shaped tumour with a central horny plug or a horny cap It is rarely more than a centimetre in diameter and is often surrounded by a ring of erythema If left alone spon taneous involution occurs and it drops off leaving a shallow depression

Squamous celled carcinoma may arise in long existing psoriasi form patches of intra epidermal carcinoma which may be multiple in patients who have been treated years previously with arsenic over long periods

The early diagnosis of secondary carcinomatous deposits in the skin is not of importance here but Paget s disease of the nipple must be mentioned It presents as a moist area slightly resembling eczema but well demarcated and indurated Later the nipple becomes eroded and eventually destroyed The condi tion indicates the presence of carcinoma in the ducts and mastectomy is necessary

Rodent ulcer or basal celled carcinoma is most commonly found on the face is slow in growth and may be multiple It may appear in a variety of forms—cystic pigmented shallow or card like ulcerating or cicatrizing These have however certain features in common The early lesion is a firm pearly nodule The margin remains well defined and has a beaded appearance often with fine vessels running over it If there is a crusted centre and this be removed the translucent appearance of the ulcer becomes manifest Only rarely does ulceration extend deeply to involve cartilage and bone and lymph nodes are never affected

Malignant melanoma is rare but because early diagnosis is so essential if the patient s life is to be saved it seems justifiable to consider it here The condition may arise in a pigmented naevus or mole and an increase in size or the appearance of irregularity *in the previously smooth surface of such a lesion* should at once arouse suspicion Sometimes a malignant melanoma first appears as a black speck in normal skin The face the sole of the foot

and nail bed are (other than the eyeball) the usual sites for this condition which is a firm tumour not invariably pigmented and which soon becomes eroded or ulcerated Enlargement of lymph nodes occurs early and tiny pigmented satellites may be seen around the melanoma On no account should the diagnosis be confirmed by biopsy as this makes the prognosis already grave even more serious Wide local excision with histological examination is the first step

ECZEMATOUS DERMATITIS

Amongst the functional dermatoses eczematous dermatitis is one of the commonest and its early diagnosis is important for if the cause can be discovered and removed and prompt treatment instituted months of disability and absence from work may be avoided *The longer the condition is allowed to persist the more* depressed does the patient become Financial problems may become acute anxiety about future prospects of employment may become intense and sometimes medico legal issues arise which *exacerbate the general tension This state of affairs is likely* to make eczematous dermatitis worse whatever may have been its original cause

Except in a court of law there is no difference between eczema and dermatitis The latter is a general term which should logically cover a wide variety of conditions including boils impetigo scalds and sunburn As the eczema reaction is one particular pattern of dermatitis it is best to call it eczematous dermatitis though often rather loosely the prefix is discarded

The primary eruption consists of pin head sized macules papules and vesicles with much itching or burning Sometimes erythema is a prominent feature sometimes oedema (as when the eyelids are affected) or sometimes large vesicles (as in pompholyx involving the thick skin of the palms and soles) In the acute phase weeping is common and secondary infection with pustules and crusting may occur Excoriations are usually seen and later in the more chronic phase there is scaling and the skin becomes thickened or lichenified as the result of friction

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It is not usually difficult to make the diagnosis of eczematous dermatitis (though occasionally psoriasis or lichen planus may

cause some doubt) but it is often far from easy to determine the cause which is an essential part of the diagnosis for until this can be eradicated the condition is not likely to resolve

Although one or other may be predominant there are usually two main aetiological factors—internal (or constitutional) and external The constitutional factors include heredity (for these conditions are often familial) age (for they are more common during infancy puberty the menopause and senescence) and deterioration in health either physical or mental It is for example well recognized that mental stress may precipitate an attack of eczematous dermatitis

The external factor may be a primary irritant substance or a sensitizing agent (allergen) Acids alkalies soap detergents turpentine mustard oil etc are by nature skin irritants which would produce eczematous dermatitis in any individual in contact with them for long enough and in a sufficient concentra tion Not so the allergens which only affect that small proportion of individuals who have become allergic to the particular sub stance previously innocuous Certain substances are especially prone to cause allergic sensitization when brought into contact with the skin and some of these are commonly used in medical practice Sulphonamides penicillin streptomycin chloramphenicol local anaesthetics and the antihistaminic drugs are examples It is best for these never to be used as skin applications or certainly not for more than a few days Once sensitization has occurred it is difficult and often impossible to desensitize the epidermis Cross sensitization may occur sensi tization to a substance may produce sensitization to a number of other chemically related compounds For example sensitization to sulphonamides may result in a similar reaction to aniline dyes such as gentian violet and acriflavine and to other substances such as benzocaine and promethazine hydrochloride (phen ergan)

Nickel dermatitis is a good example of the allergic type A woman who may have worn nickel plated suspender buckles for years without trouble becomes allergic to them probably on account of internal factors Eczematous dermatitis develops at the site of contact with nickel and the whole epidermis becomes sensitized so that a nickel plated watch or ear clips or necklace

clasp or brassiere buckles result in eczema wherever contact has been made If one of Queen Elizabeth II s sixpences (which contain nickel) be strapped to the arm for twenty four hours an area of eczematous dermatitis appears and constitutes a positive patch test The eruption on the thighs or elsewhere will not disappear until all contact with nickel is discontinued If the patient persists in wearing nickel next to the skin eczematous eruptions may appear at the sites not in contact with nickel (*e g* the forearm and eyelids) presumably because nickel is absorbed from the site of contact and carried elsewhere in the blood stream

Eczematous dermatitis resulting from contact with an allergen is usually acute (as in primula dermatitis or that from para phenyline diamine hair dyes) but it settles rapidly after removal of the cause When the face is affected there is much oedema of the eyelids and the diagnosis of angio neurotic oedema or of erysipelas may be considered The degree of contact with the allergen may be minimal though the reaction be intense On the other hand primary irritant substances usually produce at first merely a slight eczematous reaction which may become severe only after repeated exposure It may then be very slow to clear even after removal of the cause

In certain forms of eczematous dermatitis such as infantile eczema nummular eczema and pompholyx external factors play little or no part. The limb flexures and the palms and soles are often affected in eczema of constitutional origin. Eczematous dermatitis of the hands from external causes is more likely to affect the more delicate skin of the back of the hand than the tough skin of the palm The distribution of the eruption may readily indicate the external cause as in suspender dermatitis or that resulting in a susceptible person from sitting on a recently varnished lavatory seat but often the determination of the agent responsible is a very difficult detective problem In eczematous dermatitis produced by a sensitizing agent, patch tests may be useful to establish the cause when there are several possibilities as for example in dermatitis of the face thought to be caused by one or more of several cosmetic substances which have been used Patch tests must never be carried out with primary irritant substances unless they have been so diluted that the irritant

effect has been lost and any eczematous reaction to the test material can be ascribed to allergic sensitization Substances which are not primary irritants should be tested in the strength in which they are usually handled whether in the form of solutions (*e g* penicillin solution antiseptic solution) or solids (*e g* primula leaf or flower) or of emulsions (*e g* local anaesthetic cream)

Industrial dermatitis is not different from other forms of eczematous dermatitis except in its relationship to occupation Its diagnosis is often no easy problem and one which frequently faces the general practitioner in certain areas If he reaches the conclusion that an eruption is constitutional and unrelated to the man s employment he certifies that it is eczema and the patient not receiving industrial benefit sometimes feels aggrieved If the doctor thinks that it is caused by the man s employment or that he should be given the benefit of any doubt he certifies that the patient has dermatitis and the machinery of the Ministry of National Insurance is brought into operation Examination by a dermatologist may result in confirmation of the diagnosis but if not and if the man s claim for industrial benefit is rejected he believes himself unjustly treated All this does nothing but harm to a patient with constitutional eczema unless he feels that his case has been most carefully considered

Eczematous dermatitis may arise in those working in industry in several ways —

1 From contact with primary irritant substances which are usually liquids or dusts and which affect most of those at risk

2 From contact with a substance to which the worker has become allergic This may be a liquid dust or vapour and very few of those at risk will be affected

3 From exposure of an individual prone to constitutional eczema to conditions which may provoke or aggravate the eczema Such an individual may suffer from a relapse of constitutional eczema because his work is unsuitable for him—or merely because he dislikes the work or has had a row with his foreman

4 After long continued wear and tear of the skin exposure to mildly irritant dusts or liquids may eventually cause eczematous dermatitis Sometimes the resistance of the skin is lowered

by friction or work in wet conditions (as in some miners) or by excessive sweating

It will be readily appreciated that the diagnosis of industrial dermatitis may present an extremely difficult problem but one which requires early solution if the patient is to be benefited

NEURODERMATITIS

The term neurodermatitis has been used rather indiscriminately during recent years to indicate any form of dermatitis especially eczematous dermatitis thought to be of nervous origin Here only the condition known by the older term of circumscribed *neurodermatitis or lichen simplex chronicus will be described* Once it is well established the cure of this disorder is difficult and it is chiefly for this reason that early diagnosis is of practical importance

Lichen simplex is a chronic localized thickening of the skin *which results* from rubbing or scratching and *it occurs in adults* more commonly in women than men An area of apparently normal skin becomes intensely itchy and because of the repeated friction thickening or lichenification appears A notable feature is that the irritation is primary and that the skin changes occur later though occasionally neurodermatitis appears at the site of some minor organic lesion (such as a wart or a septic spot) which is frequently fingered

The intermittent pruritus is often provoked by touching the area or by a sudden emotional upset It increases to a crisis accompanied by furious rubbing or scratching until a kind of orgasm is achieved Little by little lichenification appears and eventually there is a well defined slightly raised plaque often oval in shape and several centimetres in breadth depending on the site affected The surface is leathery dry and sometimes a little scaly In colour it is reddish purple with varying degrees of pigmentation especially around the margin

Almost any part of the surface of the body may be affected but varying from person to person it is seen more commonly in certain sites the nape and sides of the neck the eyelids the points of the elbows and either aspect of the forearms the inner and upper part of the thighs the front of the knees the legs and

effect has been lost and any eczematous reaction to the test material can be ascribed to allergic sensitization Substances which are not primary irritants should be tested in the strength in which they are usually handled whether in the form of solutions (e g penicillin solution antiseptic solution) or solids (e g primula leaf or flower) or of emulsions (e g local anaesthetic cream)

Industrial dermatitis is not different from other forms of eczematous dermatitis except in its relationship to occupation Its diagnosis is often no easy problem and one which frequently faces the general practitioner in certain areas If he reaches the conclusion that an eruption is constitutional and unrelated to the man s employment he certifies that it is eczema and the patient not receiving industrial benefit sometimes feels aggrieved If the doctor thinks that it is caused by the man s employment or that he should be given the benefit of any doubt he certifies that the patient has dermatitis and the machinery of the Ministry of National Insurance is brought into operation Examination by a dermatologist may result in confirmation of the diagnosis but if not and if the man s claim for industrial benefit is rejected he believes himself unjustly treated All this does nothing but harm to a patient with constitutional eczema unless he feels that his case has been most carefully considered

Eczematous dermatitis may arise in those working in industry in several ways —

1 From contact with primary irritant substances which are usually liquids or dusts and which affect most of those at risk

2 From contact with a substance to which the worker has become allergic This may be a liquid dust or vapour and very few of those at risk will be affected

3 From exposure of an individual prone to constitutional eczema to conditions which may provoke or aggravate the eczema Such an individual may suffer from a relapse of constitutional eczema because his work is unsuitable for him—or merely because he dislikes the work or has had a row with his foreman

4 After long continued wear and tear of the skin exposure to mildly irritant dusts or liquids may eventually cause eczematous dermatitis Sometimes the resistance of the skin is lowered

The feet should be examined for tinea pedis and scrapings from the nails examined microscopically and by culture for fungus if there is any doubt A family history of psoriasis provides a valuable clue

More commonly psoriasis appears on the points of the elbows the front of the knees and on the trunk with the distinctive well defined patches covered by thick silvery scales and varying in size from a few millimetres in diameter to large plaques Psoriasis is much more common on the face than one would gather from most text books Itching is unusual except in flexural psoriasis In children the onset is often marked by a profuse eruption of guttate lesions on the trunk and limbs after an upper respiratory tract infection particularly tonsillitis

Flexural psoriasis can provide a diagnostic difficulty for those not familiar with the condition It is most common in women about the time of the menopause but also occurs in middle aged men particularly the obese Smooth red well demarcated areas may appear in the natal cleft around the vulva in the crutch and groins the axillae and under the breasts There is often fissuring which causes a certain amount of pain but itching is the most troublesome symptom and for this reason flexural psoriasis is often mistaken for an eczematous eruption An accurate diagnosis is important because flexural psoriasis can be cleared up so readily with dithranol gr ¼ in one ounce of Lassar s paste

LICHEN PLANUS

The primary lesion of lichen planus is an itchy flat topped papule which is seen to have a shiny surface when observed with light reflected from it The violet colour of the eruption is distinctive The flexor aspect of the forearms near the wrists is commonly affected and so is the trunk around the waist line In that situation the papules may be profuse and tiny and may give rise to the mistaken diagnosis of eczematous dermatitis but on careful examination the papules are seen to be flat topped larger more typical papules will probably be found elsewhere On the legs lichen planus is deeper in colour and often a little warty Lesions are not uncommon on the male genitalia and in this situation they are often annular The mouth should always be

ankles the buttocks the pubis the ano genital region and over the scapulae The finger nails are usually brightly polished and clothing over the lichenified plaque may show signs of wear

Although a distinct dermatosis lichen simplex is merely one of the many ways in which a human being may respond to the slings and arrows of outrageous fortune Patients with this condition are tense and anxious Itching and rubbing of the affected area of skin occurs when they are excited frustrated mentally upset or irritated or simply concentrating on a problem The cause of the emotional tension may be related to home life or occupation or both These individuals take life seriously and find relaxation difficult—even being obliged for some reason to sit still may provoke the irritation

It is not difficult to diagnose this condition but often far from easy to find the precipitating cause In the differential diagnosis psoriasis and lichen planus may require consideration but the former is not often itchy and in both conditions typical lesions will usually be found elsewhere The absence of previous vesicu lation is evidence against the diagnosis of eczematous dermatitis which is usually not so well circumscribed as neurodermatitis

It is essential that the patient should appreciate the nature of the condition and gain some insight into its cause This can only be achieved after a detailed history has been taken the patient being given every opportunity to talk and after a careful physical examination This is important because these patients are often highly critical They have no confidence in a doctor whom they do not consider to be thorough and the often unspoken fear of an organic disease such as cancer must if possible be relieved Moreover in neurodermatitis there may be a background of physical disease

PSORIASIS

The early diagnosis of psoriasis is not likely to cause difficulty unless it is localized affecting for example only the scalp or the nails But the lesions in the scalp are quite distinctive being thick and well defined Affected nails are dystrophic and often pitted and usually but not invariably many nails are affected The possibility of ring worm infection may require consideration

patients and though complications resulting from the use of drugs whether skin eruptions (dermatitis medicamentosa) blood diseases or other troubles are fortunately rare they are never theless serious and if the therapy be continued life may be endangered Early diagnosis is therefore of the greatest importance

Reactions to drugs absorbed internally may be either toxic or allergic though it may not always be possible to distinguish the two From the pathological aspect the acute primary lesion is in the smaller blood vessels and results in a variety of manifesta tions such as erythema purpura urticaria or nodular lesions (erythema nodosum polyarteritis nodosa) Sometimes gangrene may occur Occasionally the picture is one of anaphylactic shock

Every pattern of dermatosis may be provoked by drugs Some of them such as that resembling lichen planus which may follow treatment with gold or mepacrine may be chronic lasting for years after the drug was taken Some only appear several decades after treatment as may be seen with basal or squamous celled carcinoma following arsenical therapy

The acute types of dermatitis medicamentosa are characterized by their sudden onset the symmetrical distribution of the eruption which increases in intensity if treatment be continued and frequently by the presence of fever It may be possible to deter mine from the history which drugs have been taken but the patient may forget having swallowed various proprietary pills or powders for headache fever or constipation or may withhold the information Although dermatitis medicamentosa may be the chief manifestation the skin is not likely to be affected alone and a variety of symptoms referable to other systems may be present—renal failure asthma joint pains blood dyscrasias gastro intestinal upsets or mental changes such as confusion or delirium Unless the drug responsible is withdrawn death may occur It is impossible here to do more than to survey briefly some of the eruptions which may be seen and to mention a few of the drugs which may cause them The subject is so large that a text book of dermatology must be consulted for details Although certain drugs may produce a dermatosis with a pattern which is quite distinctive and diagnostic (*e g* carbromal and

examined as in lichen planus the buccal mucosa is affected in about half the cases Irregular white patches which look a little like porcelain may be seen both on the inside of the cheeks and on the lips and tongue Lichen planus in the mouth is usually symptomless

URTICARIA

The evanescent weals and intense itching of urticaria are readily identified but it is often difficult to find the cause Occa sionally it may be found that some particular food is responsible or that the condition follows taking a drug such as aspirin by mouth or an injection of serum or penicillin The giving of enemas and infestation with intestinal parasites are uncommon causes nowadays In chronic urticaria all these must be con sidered though there seems little doubt that in a large proportion of patients the condition is psychogenic

Papular urticaria of childhood (heat spots or strophulus) may result from some emotional upset in a highly strung child but it often clears up when the intake of sweets and ice cream is reduced and it may be associated with some particular food such as porridge bananas or chocolate However the possibility of flea bites must be kept in mind especially fleas from cats or dogs

DRUG ERUPTIONS

Eruptions resulting from the external use of drugs have been briefly mentioned in the discussion on eczematous dermatitis Here we must deal with skin disorders caused by drugs taken internally whether taken by mouth given by injection absorbed through the skin by inunction inhaled or inserted into some cavity as sulphonamide or penicillin is sometimes inserted after extraction of teeth

An eminent Victorian physician said to his students We may or we may not be able to do this patient some good but for God s sake gentlemen let us do her no harm This adjuration is still not out of place in an era of medicine notable for a wealth of new drugs Antibiotics antihistamines new hormones new analgesics and many others are examples of more and more powerful drugs in general use We can only too easily harm our

light is a rare complication of treatment with sulphonamide or gold

Several drugs in common use are liable to provoke *urticaria* aspirin and penicillin (oral or intramuscular) are examples There is often a latent period of ten to twenty days following penicillin injections before the rash appears Urticaria provoked by drugs indicates dermal allergic sensitization with the release of histamine It is *the only form of dermatitis medicamentosa in which treatment with antihistamine drugs is effective* The eruption may be severe and extensive Itching is intense Weals may be large and the appearance may be that of angioneurotic oedema

Little will be said here about the *nodular lesions* resulting from drugs Sulphathiazole occasionally provokes an eruption on the shins and forearms similar to erythema nodosum and I have seen recurrent erythema nodosum caused by tab codeine co but as the patient would not agree to be tested with the individual constituents the one responsible is not known Polyarteritis nodosa has been shown in a number of instances to be related to sensitization to sulphonamides

A curious condition occasionally seen is that in which sensitization to a drug is *limited to a well defined area or areas of skin* Each time the drug is taken the sensitized area becomes erythematous oedematous and irritable the plaque slowly disappearing once the drug has been discontinued but leaving pigmentation which may persist for months The colour of the plaques is often violaceous especially when provoked by phenol phthalein—which is the commonest cause of this condition It is contained in a variety of preparations such as paraffin agar emulsions some toothpastes and chewing gums laxative chocolate and many purgative tablets Phenacetin phenazone (antipyrine) quinine and aspirin may also produce these fixed drug eruptions

A variety of drugs including bromides and iodides may cause *vesicular* and *bullous eruptions* The vesicles often become pustular but are usually sterile The eruption may be acneiform and the pustules are rather firm to the touch With both iodide and bromide eruptions the lesions may become granulomatous and fungating

bromide) many such as the sulphonamides can produce a great variety of eruptions

Erythema may dominate the picture Erythema multiforme is only rarely provoked by drugs (*e g* barbiturates sulphonamides mysoline) but erosions of the lips tongue and buccal mucosa may be seen as in the Stevens Johnson syndrome Quinine sulphonamide and phenobarbitone are among the drugs which sometimes produce a scarlatiniform eruption often followed by desquamation Alternatively drug eruptions may be morbilli form or may show a punctate erythema resembling rubella With all these varieties of erythema fever is common Erythema of the palms and soles usually accompanied by itching occasionally appears a week or two after the intramuscular injection of penicillin

Erythematous drug reactions may be associated with *purpura* indicating not merely dilatation of capillaries but damage to their walls On the other hand the purpura may be thrombocytopenic, as when it follows treatment with gold or sedormid Purpura resulting from capillary damage is more common in the legs where its early appearance is no doubt due to the higher capillary pressure which results from an upright posture

The distinctive purpuric eruption caused by carbromal is being seen with increasing frequency Itchy patches are found with erythema petechiae purpuric streaks and pigmentation covered by fine scaling It is usually most profuse on the legs and is often confluent there especially when varicose veins and slight oedema are present but it may be seen scattered over the trunk and upper limbs On stopping the drug the rash may fade in a few weeks but sometimes persists for months

If local treatment of the skin with a drug has provoked epi dermal sensitization in the past the *eczematous dermatitis* will reappear when the same drug is given internally The area of skin previously treated is likely to be first affected though the eruption may become widespread Thus dermatitis of the eye lids may have occurred during treatment with sulphacetamide drops Later treatment with sulphonamide internally will cause a relapse of the eruption on the eyelids followed by a rash else where perhaps on the limbs and trunk and in males especially on the genitalia Actinic dermatitis from sensitization to sun

dryness and atrophy of the skin it may be a symptom of loneli ness and lack of outside interests In such old patients intro duction to old peoples clubs and other activities may greatly help

Generalized pruritus may also be a symptom of some metabolic upset It is common in obstructive jaundice and easily diagnosed but it is very rare in diabetics in whom localized anogenital pruritus occurs Uraemia is another rare cause Although un common the reticuloses should be remembered as a cause of generalized pruritus which may long precede other manifestations of the disease The writer has seen one patient who suffered from generalized irritation for seven years before the first palpable lymph node and other signs of Hodgkin s disease appeared Occasionally generalized pruritus occurs with carcinoma of an internal organ It is a symptom which must always be approached as a general medical problem

Any brief discussion of the early diagnosis of skin disorders is inevitably incomplete It is chiefly important that the prin ciples of diagnosis should be grasped the importance of the history the general assessment of the patient the determination of the primary lesion the decision as to whether the condition is a pathological affair or a disorder of function We all make blunders in diagnosis and these usually result from an incomplete examination of the patient In a busy surgery and with a fully dressed patient this is not easily avoided When there is any doubt the patient should always be examined in bed

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The problem of dermatitis medicamentosa may appear rather formidable but real difficulty in diagnosis only arises when the condition is not borne in mind and when an accurate history cannot be obtained

ITCHING

Itching is such a common symptom that its significance must be discussed however briefly When irritation results from external agents such as insects or the wearing of wool next to the skin the cause is usually obvious though scabies which is becoming more common can be missed if not kept in mind Using a lens burrows should be searched for on the palms and the sides of the fingers the wrists the anterior axillary folds the breasts in women the buttocks the genitals in males the ankles and the soles In infants vesicles or pustules may be seen on palms and soles and at any age secondary infection may lead to multiple boils on the buttocks All individuals in contact with the patient should be examined but it must be remembered that there may not be any itching during the first three or four weeks of infestation with mites The diagnosis is confirmed if a mite can be demonstrated microscopically The burrow should be gently scratched with the point of a needle where the mite is seen at one end as a dot The mite readily sticks to the needle point and can be mounted on a glass slide

Certain skin disorders associated with irritation such as eczematous dermatitis lichen simplex urticaria and lichen planus have already been discussed Anogenital pruritus requires consideration Although glycosuria candidiasis (monilia infection) antibiotic therapy thread worms organic disease of the rectum vaginal discharge and a spread of fungous infection from tinea pedis must all be eliminated as possible causes the condition in the majority of patients with anogenital pruritus is psychogenic and this aspect should be discussed with the patient who is often of the tense obsessional type

Generalized pruritus with no skin disorder apart from secondary excoriations may provide a difficult diagnostic problem At any age it may be a symptom of psychiatric disorder a mild anxiety state or more serious conditions such as depressive psychosis and parasitophobia In the elderly associated with

the number of new industrial hazards particularly in the chemical field is rapidly increasing

Finally there is much to be gained if the practitioner can see the actual working conditions when the history suggests that they may be harmful First hand accurate information may thus be obtained *concerning substances about the nature of which the* patient may be ignorant although he is handling them daily or that he may have been warned against but treats with a contempt bred of long familiarity

TOXIC CHEMICALS

Although there are a large number of new chemicals used in industry which are actually or potentially dangerous poisoning still often occurs from familiar substances such as lead or benzol **Lead poisoning** is often overlooked since the earliest symptoms are tiredness irritability and intestinal colic which if mild may be ascribed to dietary causes Even when their cause is recognized in a lead worker medical advice may not be sought and a man may quietly change to another process for a few months if he develops these symptoms The colic which usually follows several days of constipation may be associated with vomiting and is typically spasmodic It is sometimes severe enough to suggest at first an acute abdominal emergency such as appendicitis or a perforated ulcer and the abdomen may be tense and scaphoid A surgical opinion may be necessary to help in distinguishing between lead colic and an acute abdomen in a lead worker Pallor and a pinched expression are characteristic of lead poisoning and a mild hypochromic anaemia is usual This is rarely as low as sixty per cent haemoglobin and a figure lower than this is unlikely to be due to lead Where the teeth are bad and oral hygiene poor the classical blue grey particles of the lead line may be seen near the gum margin and should be sought in good light with a hand lens The line is not found in the edentulous and must be distinguished from the more diffuse bluish discoloration of pyorrhoea which extends right up to the gum margin

Anaemia and a lead line are confirmatory evidence of poisoning but of themselves indicate only lead absorption Colic is the

CHAPTER XXV

COMMON OCCUPATIONAL DISEASES

By R I McCallum

As in other fields of medicine it is often difficult to assess the significance of early symptoms or signs of disease due to occupation or to determine whether actual disease is present or not This situation is complicated rather than simplified by the use of special chemical or radiological examinations to detect exposure to a hazard Thus for example the earliest radiological change in pneumoconiosis of coal miners is not considered to be a disease and lead absorption as shown by chemical tests on urine or on blood examination is not synonymous with lead poisoning even though an abnormal amount of lead may be present in the body The detection of abnormality before manifest disease develops is of paramount importance in the prevention of occupational diseases through control both of the environment and of the human element in industrial processes In hazardous occupations where routine examinations are made of the blood or urine for toxic substances or their effects early diagnosis means *diagnosis of exposure* rather than of disease and the problem is then to decide whether the exposure is harmful or excessive

Occupational disease must be considered in the differential diagnosis of so many medical and surgical conditions that an adequate occupational history is an essential feature of full clinical assessment This should include not only information about present work sufficient to present a clear picture of what the patient actually does and the substances he handles or is likely to be exposed to but it should also give a detailed picture of all previous work from the time he left school Exposure to harmful materials may lead to disease many years later when the circumstances have been forgotten by the patient The routine occupational history is all the more important at the present time when

either stimulation or depression of the bone marrow and therefore very variable changes in the peripheral blood For this reason any abnormal finding in the blood of persons coming in contact with benzol should be considered serious and a full investigation made This should include a complete blood count with differential white cell count and probably also examination of bone marrow If possible measurement of the amount of benzol in the air at the place of work should be made as this may help to resolve any doubts about an equivocal blood picture

Clinically there are at first vague symptoms of fatigue weakness nausea loss of weight and anorexia and pallor Examination of the blood may show an anaemia perhaps with leucopenia and a reticulocytosis but some patients present with aplastic anaemia or leukaemia Recognition of aetiology will depend on the occupational history and proof of exposure Benzol poisoning must always be considered in the differential diagnosis of these blood conditions particularly as there may be a delay in onset of up to two years after exposure has ceased It may then of course be impossible to obtain appropriate environmental measurements and direct proof may be lacking It appears that some individuals are particularly susceptible to benzol poisoning and once there is reasonable evidence of blood changes they should not return to work entailing possible exposure

Men working in the rubber and textile industries or in the manufacture of dyes and inks may come into contact with **aniline,** a colourless or yellow brown oily liquid Inhalation of its vapour or splashing on the skin (from which it is easily absorbed) may lead to poisoning the most striking features of which are blueness of the face and giddiness The blue discoloration of the skin (which is due to methaemoglobinaemia) occurs particularly in hot weather and it is best seen in good natural rather than artificial light If absorption continues from extensive skin splashes or contaminated clothing then there is a danger of severe prostration and death in coma Continued mild exposure may lead to some degree of anaemia insomnia or angina The blue appearance with the history of exposure should make the diagnosis straightforward At one time dye workers were thought

earliest definite sign of actual poisoning whilst wrist drop or foot drop are uncommon and encephalopathy very rare The work history will often provide the diagnosis if it is remembered that lead is encountered in widely different industries and pro cesses apart from the lead industry itself and that the common mode of entry into the body is by inhalation of lead containing dust or fumes For example lead fumes may be inhaled during burning through painted steel plates in shipbreaking or from the burning of waste paper from battery works in completely uncontrolled conditions or dust may be inhaled by painters dry sandpapering painted wood Laboratory investigations such as the stipple cell (punctate basophil) count reticulocyte count and the estimation of lead and coproporphyrin excretion in the urine are very helpful and should be done if possible but unless they are grossly raised they too usually only indicate lead absorption and do not closely parallel clinical lead poisoning in the individual

Negative results are also valuable a thirty one year old labourer in a red lead process had several attacks of colic and both he and his doctor were convinced that he had lead poisoning His haemoglobin was normal however there were no punctate basophils in his blood and the urinary coproporphyrins were within the normal range On further enquiry it was found that his youngest child was under hospital treatment for enteritis *Salmonella typhimurium* was isolated from the man s faeces and was almost certainly the cause of his symptoms

The treatment of lead poisoning is now highly effective since the introduction of the chelating agent calcium disodium E D T A (ethylenediamine tetra acetate) which is usually given by intra venous infusion

Exposure to **benzol** (benzene) may occur in gas works and in the distillation of coal tar Benzol is used very widely in the chemical industry as a solvent in rubber glue paint and varnish manufacture and in the aeroplane industry It must be distinguished from *benzine* which is a quite different chemical of the paraffin series Acute poisoning from inhalation of the highly volatile benzol vapour produces excitement, headache paraes thesiae in the extremities and fatal coma if exposure is pro longed Chronic poisoning from continuous daily exposure to small quantities of benzol vapour is the greater danger and is much less dramatic and more difficult to control This causes

nervous system to produce tremor ataxia and constriction of the visual fields

The mental effects of chronic mercury poisoning are known as erethism (from the Greek word for irritation) which refers to the ease with which the patient is upset He is embarrassed and timid with strangers lacks confidence and suffers from loss of memory and depression Marked intention tremor of the upper limbs may be present and the handwriting appears shaky and indistinct Fine tremor of the orbital muscles and of the lips and tongue with spells of coarser movements also occur Stomatitis may appear early during exposure with excessive salivation and tender swollen gums causing pain on eating but a mercurial line on the gums is rare It may be difficult to distinguish erethism from a mild psychoneurotic depression while the tremor must be differentiated from that due to nervousness senility paralysis agitans hyperthyroidism or disseminated sclerosis The combination of emotional upset with tremor and gingivitis where there is a probable mercury hazard should leave no doubt as to the diagnosis Estimation of mercury in the urine may confirm that there has been excessive exposure

A fitter aged forty years employed in maintaining plant in a mercury battery factory complained of tiredness dizziness headache depression and suicidal thoughts His hands trembled and he tended to burst into tears easily These symptoms developed over a period of two or three months during which he worked overtime averaging sixty five to seventy hours work a week He was found to have 680 micrograms of mercury in a twenty four hour specimen of urine about seven times the upper limit of normal

As agriculture becomes more scientific and mechanized so the risks to the agricultural worker alter in character Toxic chemicals are now used for the control of weeds and insect pests during several months from April onwards particularly in spells of good weather These chemicals can affect not only those engaged in their manufacture but also spraying contractors and their employees farm workers and workers in greenhouses orchards hop fields market gardens and seed dressing Although most of the large number of different chemicals in common use are comparatively safe vague symptoms in an agricultural worker should raise the possibility of contact with a toxic chemical Absorption may be from skin splashes with

to contract bladder carcinoma from aniline but this is now known to be incorrect and the causative agents have been identified as β naphthylamine or benzidine

Carbon tetrachloride is familiar as a household cleaning fluid under proprietary names It is a colourless solvent smelling not unlike chloroform with the great advantage of being non inflammable It is widely used in dry cleaning as a fire extinguisher as an anthelmintic and in the rubber chemical and paint industries It may sometimes be taken by mouth in mistake for water but poisoning is most likely to occur from exposure to its vapour in a confined atmosphere Its main effect is narcotic but it can also cause kidney and liver damage The characteristic symptoms of chronic tetrachloride poisoning are mental confusion and gastro intestinal upset There are no physical signs but the patient complains of light headed ness giddiness and headache There may be alteration in sleep rhythm with drowsiness by day and insomnia suggesting a psychoneurosis Recurrent nausea and vomiting colic of variable severity and diarrhoea with vague abdominal discomfort and a perverted sense of taste and smell contribute to an indefinite picture which may be confused with a primarily gastro intestinal disease Unconsciousness follows acute exposure to a high con centration of carbon tetrachloride and on recovery from this jaundice and enlargement of the liver with oliguria and uraemia and oedema of the lungs may follow There is much individual variation in susceptibility to carbon tetrachloride and there may be no serious results in some men after heavy exposure

Mercury poisoning in industry is mainly chronic and presents as a combination of psychological upset tremors and stomatitis It has long been associated with the felt hat industry in which mercuric nitrate is used, but nowadays it is likely to arise in the manufacture of thermometers and barometers of electrical contacts and lamps and of explosives and fungicides It may be inhaled as a dust of mercury salts or as a vapour metallic mercury being volatile at ordinary temperatures Poisoning has been described in police officers using a powder of mercury and chalk to show up fingerprints for photography Organic mercury compounds such as those used in fungicides act on the central

exposed to organic phosphorus insecticide should be presumed evidence of poisoning until proved otherwise As these sub stances are absorbed through the skin clothing should be promptly removed and the body washed

It is vitally important to know accurately what material has been used for the emergency treatment of organic phosphorus poison ing is to give atropine sulphate in doses as high as one to two mg intramuscularly or intravenously and to repeat them frequently for a prolonged period depending on the severity of the case In D N O C poisoning on the other hand this treatment is absolutely contra indicated and may cause a fatality Labora tory confirmation of organic phosphorus poisoning is obtained by demonstrating reduced cholinesterase activity in the blood

OCCUPATIONAL INFECTIONS

Cutaneous anthrax, the malignant pustule is still a risk for men who come in contact with animals or animal products such as hairs skins or horns Butchers tanners slaughtermen wool and hair sorters brush makers and dock labourers may all handle infected material as may men in glue bone meal and gelatine factories who work with dried bones from India Anthrax begins as a small red pimple on the face or neck or sometimes on the limbs or trunk which increases in size to form a typical round black scar surrounded by small vesicles and oedema Early recognition is important as penicillin is an effective treatment and may prevent a blood stream invasion which though uncommon can be fatal Examination of exu date from the vesicles for bacilli should be made but treatment should not be delayed on this account Since anthrax is un common workers in industries in which there is a risk of infection are given individual cards to show to their doctor drawing attention to the hazard

Erysipeloid of Rosenbach is a skin infection common amongst fishermen and in the fish trade in butchers cooks and housewives who may handle decomposing animal tissue and in workers with old bones It is more common in summer and early autumn than in winter or spring It is particularly well

concentrated preparations from contaminated equipment or from prolonged inhalation of airborne mist or dust in spraying especially the latter

The most important poisons are D N O C (*dinitro ortho cresol*) and organic phosphorus compounds such as parathion and T E P P (*tetra ethyl pyro phosphate*) sprayers must wear protective clothing in all such cases Absorption of these chemicals into the body is cumulative and the early symptoms are easily overlooked by both workmen and doctor

D N O C is a selective weed killer for cereals the action of which in the body is to increase the basal metabolic rate and to interfere with carbohydrate metabolism Poisoning is most likely to occur in hot weather It may present first as excessive sweating and thirst in a man with yellow stained hands face and hair who feels extraordinarily well In more severe cases sweating is profuse there is a rapid pulse nausea and vomiting restlessness air hunger and fever Dehydration becomes marked with scanty yellow urine containing albumin and finally there are convulsions coma and death There is no specific treatment for this condition and symptomatic measures to cool the body relieve anoxia and counteract dehydration must be used

The **organic phosphorus compounds** such as parathion are insecticides particularly for use against aphides and red spider In man they destroy the enzyme cholinesterase so that excess acetylcholine accumulates at nerve endings The effects are therefore those of powerful parasympathetic stimulation and can be grouped as —

1 Secretory diarrhoea sweating and salivation bronchial constriction and pulmonary oedema Nausea and abdominal pain also occur

2 Effects on the central nervous system such as headache restlessness ataxia pin point pupils tremors and convulsions and

3 Twitching and weakness of muscles

Some but not all of these symptoms may be seen in any one patient The presenting symptoms depend on the properties of the particular poison and its route of absorption so that at first they may be predominantly gastro intestinal pulmonary or nervous Any of these symptoms in a person who has been

of Weil s disease in which pain vomiting and rigidity may suggest an acute abdomen or—if there is haematemesis—a bleeding gastric ulcer Occasionally the symptoms are predominantly respiratory with pneumonia and haemoptysis and in others there are symptoms suggesting meningitis The jaundice must be differentiated from infective hepatitis in which the onset is less abrupt and less severe As the name of the organism suggests there may be haemorrhages into skin mucous membranes or various organs as part of the clinical picture

The diagnosis is confirmed by the appearance of agglutinins in the blood after five or six days and their rapid increase in the second to third week of the illness The organism itself may be found in the blood by culture in the first week in the urine during the second week and where meningeal signs are present in the cerebrospinal fluid

INHALED DUSTS PNEUMOCONIOSIS

Pneumoconiosis is a comprehensive term used to describe lung disease produced by inhalation of dusts of small particle size and varied biological activity over a period of months or years It is diagnosed from a radiograph of the lungs in conjunction with a detailed occupational history There is usually a long interval between exposure to dust and the development of signs and symptoms and as a result the X ray findings often relate to industrial conditions which no longer exist On the other hand too much attention paid to the occupational history may occasionally lead to misdiagnosis of one or other of the common lung or heart conditions which cause breathlessness and cough Clinical examination in pneumoconiosis gives few positive signs but serves to exclude other diseases which may simulate it

The commonest forms of pneumoconiosis are '**pneumoconiosis of coal workers**' (sometimes called anthracosis) and **silicosis** Coal workers pneumoconiosis is practically confined to coal miners who form a very large occupational group but it also occurs in a small number of trimmers handling coal in confined spaces in the holds of ships Silicosis occurs in men exposed to dust containing silica (SiO_2) in a wide range of industries such as the manufacture of refractory (fire) bricks quarry

known in certain areas such as Aberdeen where many are exposed to the risk of infection in the fish trade but it may be less familiar to doctors elsewhere It usually occurs on the fingers a few days after a minor injury such as a prick from fish scales or bone or a superficial knife cut which may pass unnoticed Erysipeloid is a pink or reddish blue swelling with a well demarcated edge slowly spreading from the point of injury It involves skin and subcutaneous tissue but there is no suppuration nor any complications Occasionally there is fever malaise giddiness and headache with a generalized ery thema and urticaria The dorsum of the hand and adjacent fingers may be involved with stiffness and pain in the interphalangeal joints The condition is self limiting and resolves spontaneously in from one to three weeks but this period may be reduced with penicillin Erysipeloid must be distinguished from a pyogenic infection such as cellulitis or whitlow Erysipeloid does not sup purate is not very tender and unless there is secondary infection is unaccompanied by lymphangitis or by gland or joint involve ment. **Erysipelas** on the other hand is a streptococcal skin infection and is more severe It may affect the face or head and can be accompanied by severe headache and high fever while at the margin of the erythema are small vesicles filled with clear or turbid fluid containing streptococci

Weil's disease is an infection with *leptospira ictero haemor rhagiae* which is spread by passage through rat urine so that men working in contaminated water are liable to contract it These include coal miners fish market men sewermen farmers canal workers butchers piggery workers and hedgers and ditchers Weil s disease may present in three forms first as a sub clinical infection as is suggested by finding antibodies to the leptospira in the blood of apparently healthy men Secondly it may appear as a mild fever without jaundice which may be regarded as influenza and thirdly as a severe infection usually with jaundice and haemorrhages The onset is sudden with fever and headache severe muscular pains prostration and vomiting and jaundice appears after about a week in two thirds of cases Albumin red cells and casts are found in the urine and the blood urea may be raised There is also an abdominal type

or pneumoconiosis specialist there may be contradictory opinions on the same films The situation is made worse by the difficulty even in the best radiological departments in obtaining chest films of suitable technical quality on which a sound opinion can be given The earliest detectable lung changes requiring first class radiological technique for their demonstration may not con stitute disease either clinically or legally and to some may seem *like beauty to lie in the eye of the beholder*

Further confusion is bound to occur because pneumoconiosis is defined legally in terms of pathology the diagnosis in life is made largely on radiological findings and the symptoms when present are not specific and do not necessarily relate to either

Asbestosis is much less common than either coal workers pneumoconiosis or silicosis and again the symptoms are non specific After ten or more years work with asbestos an insidious onset of breathlessness on exertion and a succession of chest colds pass gradually into a syndrome suggesting bronchitis or bronchi ectasis At first there may be no radiological signs and only the work history will suggest the diagnosis This may be pipe lagging or work in a factory making asbestos roofing brake linings acid resisting cloth valve packings fire fighting suits or boiler mat tresses

SKIN IRRITANTS

Industrial dermatitis occurs in practically every industry and has great importance because of the large number of workers involved and the large amount of time lost through it Both direct irritation and sensitization of the skin are produced by a wide variety of substances and the true cause of such a lesion is often obscure An occupational origin should be suspected if the lesions appear on exposed parts of the body are made worse by contact and improve on removal from exposure Further evidence in favour of an occupational origin is provided if the appearance of the rash is consistent with known cases and if other workers exposed to the same material develop similar trouble A patch test with the suspected material can be used to confirm the diagnosis but this test has limitations and should be carried out by a dermatologist As the part played by constitutional factors is uncertain it is better to avoid a firm diagnosis of occupa

ing and stone dressing pottery manufacture steel dressing and tunnelling through rock Iron ore lead and tin miners may all cut drill or blast rock containing silica and so do many coal miners who may thus contract silicosis or pneumoconiosis of coal workers or both according to the type of work done

When pneumoconiosis presents with symptoms these may suggest bronchitis and emphysema or heart disease The gradual onset of breathlessness on exertion with or without cough sputum and wheezing are symptoms common to most forms of occupational lung disease and in many patients with pneumoconiosis chronic bronchitis is obviously part of the clinical picture The bronchitis may in fact be the most disabling element in the disease but its relationship to the pneumoconiosis is far from clear The combination of these symptoms with a history of work over a period of several years in coal or mineral mining or one of the other occupations mentioned above should *suggest pneumoconiosis as a possible cause*

Symptomless pneumoconiosis may be discovered unexpectedly when the chest is X rayed for some other reason as in mass radiography for tuberculosis The diagnosis may be possible from the X ray alone but should be confirmed by taking a work history and by a thorough clinical examination to exclude other disease as far as possible

In both silicosis and coal workers pneumoconiosis it may be possible to tell from the X ray whether there is dust alone in the lungs or whether tuberculosis is present as a complication Silicosis may sometimes present as open tuberculosis with wasting and gross signs in the lungs In coal workers *pneumoconiosis* sputum positive tuberculosis is relatively uncommon and the infection tends to form large fibrotic masses in the lungs which are not detectable on physical examination Clinically the differential diagnosis lies between bronchitis and emphysema cardiac disease (which may be superimposed on long standing pneumoconiosis) advanced pulmonary tuberculosis and carcinoma of the bronchus

The radiological diagnosis of pneumoconiosis is a skilled job demanding some experience of the condition Even then it is commonly found that largely because of individual differences in interpretation whether by a hospital radiologist chest physician

The fingers of the hand supporting the tools become numb white and stiff especially in cold weather but the index finger and thumb usually escape The affected fingers then become deeply cyanosed for a variable period of time sometimes with dull pain Recovery is associated with reddening of the fingers and painful tingling Gross disability is unusual and it is seldom necessary for the man to change his job This form of Raynaud s phenomenon can easily be distinguished from that due to other causes because of the direct connection with the job and the absence of lesions in the skin or nervous system

Noise-deafness (see also Chapter XXII) which is well known in boilermakers may also occur in any occupation in which noise is prolonged and intense such as weaving machine work or aviation It develops gradually and becomes permanent after prolonged exposure Although at first ability to hear ordinary speech may not be impaired this also is affected by continued exposure and the handicap becomes severe As the early stage of deafness is for high frequencies and is not apparent to the patient it can only be detected by audiometric tests and if such a suspicion arises otological advice should be sought

The rapidly expanding use of radioactive isotopes in science and industry and the development of atomic energy make it certain that an increasing number of occupations will carry a risk of exposure to **ionising radiation** X rays and gamma radiation are used in industry to detect flaws in welds in high pressure boilers or ships plates for examination of castings and reinforced concrete and in diffraction crystallography Apart from the familiar diagnostic and therapeutic uses of X rays and radium in medicine isotopes are now used for the investigation or treatment of polycythaemia thyroid disease and brain tumours and as tracers in research work Over exposure to X and gamma rays produces burns of the skin with redness scaling loss of hair blistering and necrosis Chronic changes present in the form of ridged deformed nails skin atrophy and wart formation pigmentation telangiectases and epitheliomatous growths many years afterwards Cataract and sterility may also occur

Isotopes emitting beta radiation are used as static eliminators in thickness gauges in devices which indicate the level of fluid or solids in containers to measure wear in machinery and to

tional dermatitis in the early stages of a skin rash A period of observation will give time for exact information about sub stances used at work to be obtained

Irritants such as **chromic acid** and its salts (sodium and potas sium chromate and bichromate and ammonium bichromate) are corrosive and produce characteristic lesions in men employed in chromium plating works if the process is inadequately controlled The lesions are round clean penetrating ulcers with a hard well defined border which occur on the basis of a cut or scratch usually on hands and arms or on the legs These ulcers can take months to heal and they leave scars but are relatively painless In the same group of men ulceration and perforation of the anterior part of the nasal septum is often found from inhalation of acid fume or mist

Cutting oil used in engineering works is a common cause of irritant dermatitis affecting the forearms front of elbows and anterior aspect of thighs and legs Depending on the type of oil there is a folliculitis or furunculosis due to mechanical blockage of sebaceous glands a diffuse maculopapular rash or a dry fissured erythema which may desquamate These mineral oils may eventually lead to hyperkeratosis and malignant changes in the skin Similarly men who work over many years with tar pitch or asphalt *e g* in road laying or with anthracene or creosote are subject to acne erythema and warts on the face forearms and scrotum which may also develop into carcinoma The coal tar derivatives are the commonest cause of occupational cancer which is still a large problem in industrial medicine On the scrotum a small wart or horny projection can remain un changed for months or years but may eventually ulcerate and spread It must be differentiated from a sebaceous cyst and treated at once with radium

PHYSICAL AGENTS

The diagnosis of disease due to unusual physical conditions is often provided by the patient who is quite familiar with it in himself or fellow workers Dead hand from ***vibratory tools*** is common amongst riveters caulkers fettlers scalers or drillers

The symptoms vary from mild discomfort in the limbs (the niggles) which often pass off spontaneously or after recompression to more severe pain in joints or limbs (the bends) or in some cases paralysis or even coma and death The bends is a dull throbbing pain of gradual onset which may come at a variable time from a few minutes to a few hours after leaving the airlock in which decompression takes place The painful limb tends to be held in a semi flexed position Other symptoms with equally graphic colloquial names are the staggers due to vertigo and the chokes which is an asthma like attack of breathlessness Itching and redness of the skin may also occur

In its most severe form decompression sickness is a rapidly developing paraplegia or quadriplegia with paralysis of the respiratory muscles As this may be permanent if recompression is not immediately carried out bends or paralysis in a compressed air worker require immediate return to a compression chamber or medical lock at the works and not to a hospital Compressed air workers are supplied with labels to carry with them stating the nature of their job and giving the address of the nearest medical lock

A foreman aged forty five years with fifteen years experience in compressed air work had suffered minor attacks of bends at different times After two hours in a tunnel under a pressure of thirty four lbs per square inch he came out into normal atmospheric pressure by way of a lock not intended for use by men in only a few minutes instead of the full forty minutes necessary for proper decompression Fifteen minutes later his legs became weak and numb and he developed paralysis and loss of sensation in both legs retention of urine and incontinence of faeces Eventually there was partial recovery but he was left permanently disabled

A delayed result of compressed air work is avascular necrosis of a bone or joint such as the humerus or hip joint It may occur several months or years after exposure and cannot be distinguished from osteoarthritis due to avascular necrosis of unknown aetiology unless the work history is known

detect leaks in pipes This type of radiation may also affect the skin to produce a slowly developing burn with eventual ulceration and scarring, and can cause cataract The main danger from radiation is frequent small overdoses through carelessness defective apparatus or bad technique Accidental irradiation of the whole body occurs *only rarely* *Prevention of over exposure* in atomic energy establishments is primarily the job of health physicists but protection of the individual worker in industry depends on the careful use of film badges or ionization chambers These are worn on the person during working hours and record the dose of radiation received during that period Routine blood counts are often done but are of limited value other than to indicate a trend because of the normal variation in an individual at different times of the day or year With chronic over exposure to radiation the red cells white cells and platelets may all be depressed but especially the lymphocytes Leukaemia or aplastic anaemia may occur if exposure has been heavy By present standards there is over exposure if a worker has received more than 0 3 *roentgen* in a week or more than three *roentgen* over a period of thirteen weeks Certain radioactive substances used in industry may also have toxicological dangers apart from their radioactivity but the latter is almost always the more important

Decompression sickness or **caisson disease** is a consequence of work in compressed air during deep sea diving or the making of *foundations for bridges or tunnelling under rivers* A similar condition in reverse occurs in airmen descending from high altitudes Air pressure greater than that of the atmosphere is necessary to keep water out of the diving chamber or tunnel *and results in solution of air in the body fluids and tissues* The oxygen is used in the body in the normal way leaving nitrogen which is especially soluble in fat and is held in the body until the pressure is reduced If this is done suddenly nitrogen is *released in the body like bubbles in soda water* when the cork is removed so that to avoid this a carefully observed pro gramme of decompression must be followed In spite of strict precautions some men can be expected to suffer minor symptoms *after decompression* while any departure from carefully controlled decompression may result in a major catastrophe

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